LEONTIASIS OSSEA

A Critical Review, with Reports of Four Original Cases

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This review is based upon 149 reported cases of leontiasis ossea, together with four cases that I have investigated and treated personally.

The term leontiasis was originally applied to elephantiasis of the facial skin complicating leprosy. Virchow (1864) described a patient with thickening of the facial bones as well as the skin and called it leontiasis ossea. It is now accepted that in true leontiasis ossea there is no thickening of the skin, and Virchow's original case was probably one of congenital gigantism of the face or of neurofibromatosis. Other authors have used the term for diffuse swellings in the jaws and for localised or diffuse swellings of the cranium. For the purpose of this review leontiasis ossea is defined as a generalised homogeneous swelling of the facial bones arising usually in the first decade of life. The swelling may spread to the lower jaw and cranium. The clinical picture shows an insidious onset of facial deformity with or without proptosis, nasal obstruction and impairment of vision. Pathologically and radiologically two main types may be recognised—in one there is sclerosis of the bones, associated mainly with infection; in the other there is an osteolytic process which is mainly unconnected with infection.

CLASSIFICATION

Many classifications have been suggested. Virchow (1864) thought that the condition arose simply as an inflammatory reaction in the periosteum—a "creeping periostitis." Many classifications (Bassoe 1905, Buhl 1878, and others) have included destructive conditions such as acromegaly and gigantism as leontiasis. Others have associated it with Paget's disease. Boit (1912) suggested that it was an endosteal reaction and cited osteitis fibrosa as a prime cause. Knaggs (1923b) differentiated two types—a "Virchow" type, or "creeping periostitis," a sclerotic process usually caused by trauma or infection; and a diffuse osteitis of the medulla of the bones. Other classifications have been suggested by Reiss (1935), Kienbock (1940), and Windholz and Cutting (1945). The main objection to most of these classifications is that they have included as examples of true leontiasis ossea conditions that can be clearly differentiated; these have tended to overshadow the importance of osteitis fibrosa and of polyostotic fibrous dysplasia, the association of which with leontiasis ossea was emphasised by Eden (1939), Falconer et al. (1942) and Etter and Hurst (1943). Recently Pugh (1945) and Schlumberger (1946) have suggested the association of monostotic fibrous dysplasia with leontiasis. I have preferred to classify leontiasis ossea as follows.

TRUE LEONTIASIS OSSEA

Type 1. Virchow's type or sclerotic type.†

Type 2. Fibro-dysplastic type which includes:

- Monostotic fibrous dysplasia.
- Albright's syndrome.
- Multilocular cystic disease of the jaws.

FALSE (OR SYMPTOMATIC) LEONTIASIS OSSEA

Paget's disease.
Congenital gigantism.
Pituitary gigantism.
Craniofacial anomaly.
Benign and malignant tumours of the nose and sinuses.
Syphilitic osteoperiostitis.

† Based on a thesis for the Doctorate of Medicine in the University of Cambridge, 1951.

‡ I prefer the term "sclerotic" type, because it suggests more directly the pathological and radiological features, which are in marked contrast to those of Type 2 leontiasis.
Of the 153 cases that have been studied, I have classified ninety-seven as examples of true leontiasis ossea. Of these, forty-five were of Type 1 and fifty-two of Type 2. Forty cases were classified as false (or symptomatic) leontiasis, and sixteen did not fall into either category (Table I).

**ETIOLOGY**

**Type 1 leontiasis.** *Injury and infection*—A history of trauma or infection is obtained in a large proportion of the patients. Out of forty-five reported cases thirteen were associated

<table>
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<tr>
<th>Classification of 153 cases of leontiasis ossea</th>
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<td>(149 reported cases and four personal cases)</td>
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<table>
<thead>
<tr>
<th>True leontiasis ossea</th>
<th>97 cases</th>
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<tbody>
<tr>
<td>Type 1</td>
<td>45 (46.3 per cent)</td>
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<tr>
<td>Type 2</td>
<td>52 (53.7 per cent)</td>
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<tr>
<th>False leontiasis ossea</th>
<th>40 cases</th>
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<tr>
<td>Syphilitic osteoperiostitis</td>
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<tr>
<td>Paget’s disease</td>
<td>6</td>
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<tr>
<td>Pituitary lesions</td>
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<tr>
<td>Craniostenosis</td>
<td>7</td>
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<tr>
<td>Congenital gigantism</td>
<td>7</td>
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<tr>
<td>Tumours of maxilla</td>
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<table>
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<tr>
<th>Unclassified</th>
<th>16 cases</th>
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<td>Localised osteitis fibrosa</td>
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<tr>
<td>Chronic hyperplasia of maxilla</td>
<td>2</td>
</tr>
<tr>
<td>Dystrohondroplasia</td>
<td>1</td>
</tr>
<tr>
<td>Albers-Schönberg disease</td>
<td>1</td>
</tr>
<tr>
<td>Neurofibromatosis</td>
<td>2</td>
</tr>
<tr>
<td>Osteogenesis imperfecta with platybasia</td>
<td>1</td>
</tr>
<tr>
<td>Albright’s syndrome not showing leontiasis</td>
<td>1</td>
</tr>
<tr>
<td>Paget’s disease not showing leontiasis</td>
<td>1</td>
</tr>
<tr>
<td>Meningioma</td>
<td>2</td>
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<tr>
<td>Undiagnosed</td>
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with injury and eleven with infection. Four others developed infection after the onset of the disease. Dacrocystitis is the commonest infection, though it is not always primary. Dental infection is more obviously primary. It is known that a chronic sclerosing osteomyelitis can arise from infection or after injury, either by interference with the blood supply or by organisation of a subperiosteal haematoma. Garré’s chronic sclerosing osteomyelitis has been suggested as a cause (Freedman 1933, Windholz 1945, Pugh 1945). Knaggs (1923b) suggested that infection was the main cause of this type but he could not substantiate it. Bailey (1939), however, reported a case of leontiasis in which there was a central Brodie’s...
abscess from which a streptococcus viridans was grown. In this case infection was obviously endosteal, but in another case in this series it arose in a subperiosteal hematoma. It should be remembered, however, that although a low-grade infection—possibly initiated by trauma—may determine the onset, subsequent severe infection can be produced by obstruction of the natural ostia of the antrum and naso-lacrimal duct by the hypertrophic bone.

Familial factors—In three cases of this series familial, and probably congenital, factors were of importance. These cases were no doubt allied to Frangenheim’s (1914) "bilateral familial maxillary hyperostoses." They were mostly characterised by grotesque enlargement of the maxilla and secondary bone infection.

From Type 2—One Type 2 case changed to Type 1 after x-ray therapy. Another patient had the local sclerosis of the face typical of Type 1, whereas the vertex showed the osteoclastic areas of Type 2. This change is probably due to decreased vascularity brought about by radiation or by senile changes in the blood vessels.

Generalised osteosclerosis—There were two cases of generalised osteosclerosis in this group. One was due to Albers-Schönberg’s disease and the other was Halliday’s (1940) case which does not fall into any of the recognised varieties of this condition. Sclerosing conditions which may give rise to Type 1 leontiasis include osteopoikilie (which affects one half of the length of a bone), Engelmann’s disease (which affects the shaft), osteopetrosis or "marble bones" (which affects the bone ends) and Ler’s melorheostosis (which shows a spotted effect of the bone—"candle dripping"—at the ends).

There are also two monostotic types: the eburnising osteitis of Mott and the condensing osteitis of Sicard. In addition, chronic lymphoid or myeloid leukaemia may give rise to true sclerosis. It may be wrong pathologically to class these cases as Type 1 leontiasis. But clinically and radiographically they resemble other Type 1 cases. They also may explain the later age incidence of the condition in some patients of this group.

Type 2 leontiasis. Injury and infection—These may play a part. Out of fifty-two reported cases of Type 2 leontiasis, three were associated with trauma, and five with infection. But all of these eight cases were monostotic.

Developmental factors—Most of the congenital cases were monostotic. There were ten congenital cases of which one was polyostotic. In three familial cases the condition was mainly monostotic and was due to familial multilocular cystic disease of the jaws.

Monostotic and polyostotic fibrous dysplasia—These conditions show symptoms such as pigmentation, thyroid enlargement and large stature, which suggest an endocrine association. Many, in fact, are associated with Albright’s syndrome. But pituitary hyperfunction produces gigantism and not fibrous dysplasia. Hamann (1920), on the other hand, suggested that fibrous dysplasia is due to hypofunction of the pituitary—a syndrome of atrophic genitals and bone cysts. Most patients with fibrous dysplasia are large for their age, and the pituitary factor must be kept in mind as a possibility.

Disease of the nervous system—Some cases have shown hypothalamic symptoms such as diabetes insipidus and inverted sleep rhythm. These also occur in the Stewart-Morel syndrome caused by hyperostosis frontalis interna, which many believe to be a type of fibrous dysplasia. Other cases were associated with a syringomyelic condition of the cord. It should be remembered that platybasia can give rise to a syringomyelic condition of the cord associated with stenosis of the foramen magnum, the etiological factor. This platybasia can be caused by Paget’s disease, dyschondroplasia and fibrous dysplasia. Associated with platybasia, various types of lateral spinal tract sclerosis can occur. For these reasons I regard the neurological origins of this disease as tenuous. It is clear that they may arise mechanically as a secondary factor in many diseases of the skull in which bone softening occurs.
Primary liver disease must be mentioned because some cases were associated with icterus neonatorum. It is known that rarefaction and cortical thinning of bone may be produced in puppies who have an artificially produced obstructive jaundice.

PATHOLOGY

Gross examination shows an enlargement of the facial bones which may be associated with swelling of the jaws and cranium. There is never any thickening of the skin; when the swelling is pronounced the skin is markedly thinned. The diploe of the cranium and the outer plate are affected.

In Type 2 leontiasis the swellings are more exuberant; the bones are softer and may even show egg-shell cracking; and there is usually more pressure on surrounding structures such as the eye and the nose. The bones most commonly affected are the anterior and orbital plates of the frontal, the orbital plate of the ethmoid, the zygoma and the maxilla. In Type 1 cases the antra are frequently obliterated. The bones of the middle fossa show sclerosis in all cases, but especially in Type 2.

Histology. Type 1—From an endosteal or periosteal stimulus there is a fibrous-tissue and osteoid hyperplasia of the medulla. The periosteal new bone is formed from fibrous bone and may arise from an endosteal or periosteal focus. The general histology is that of osteoblastic and fibroblastic activity with a minimum of osteoclasts or giant cells.

A similar histological picture will be found in cases of chronic hyperplasia of the maxilla (Westmacott 1913) and in chronic sclerosing osteomyelitis (Thoma 1941).

Type 2—There is a fibrous enlargement of the medulla which expands and atrophies the cortex. The medulla is full of fibroblasts, osteoid tissue and giant cells, in varying proportions. In many cases and in all active phases no osteoid tissue will be found at all (See Case 1 and Fig. 3). In the regenerative phase, fibrous bone will be seen growing as a metaplasia between medulla and cortex.

CLINICAL FEATURES

Sex and age incidence—Type 1—There were thirty-one males and fourteen females. This male preponderance is attributed to a greater liability to trauma or infection. The average age at onset in this type was 18.4 years. Type 2—There were twenty-four males and twenty-eight females. The average age incidence was seven years (a number of congenital cases are included).

Disease incidence—According to Fairbank (1950) the skull is affected in one-third of all cases of polyostotic fibrous dysplasia and in two-thirds of the cases of Albright’s syndrome. Schlumberger (1946) found the facial bones affected in 20 per cent of sixty-seven cases of monostotic fibrous dysplasia. My own research has revealed records of ninety-seven true cases in a period of 180 years. This indicates that the condition is rare, especially if it is restricted to the classification suggested here.

Symptoms—There is a history of an insidious onset of facial swelling, noticed in early infancy. The swelling remains unilateral for some time and the child is brought up to the doctor because of facial asymmetry. At this stage there are few symptoms beyond facial pain or neuralgia. Later, at ten to fourteen years, the child complains of nasal obstruction or proptosis. Later still there may be impairment of vision, especially a restriction of the visual fields. Finally, the secondary effects produced by the large hyperostotic mass will give epiphora, sinus infection, headache and neuralgia. Headache is common and is usually a local pressure symptom. It has been suggested that headache and coma from raised intracranial pressure are common, but I believe that this is due to an erroneous diagnosis of leontiasis being made in cases of craniosenosis or menigioma. Raised intracranial pressure was proved in only six cases of true leontiasis. Rarer symptoms are dysphagia, difficulty in closing the mouth, and difficulties of speech.

Signs—If the tumour is large the skin is stretched and thinned. The swelling involves the maxilla, zygoma or frontal regions. There may be associated swellings in the cranium or
mandible. The tumour may be unilateral at first (Fig. 1), but becomes more diffuse later. The nose may sink in between two paranasal swellings. This is a particular characteristic of the false type syphilitic and of Frangenheim’s variety of Type I leontiasis. 

**Nasal signs**—Nasal stenosis is the commonest finding (twenty-two cases in this series). The block is due to pressure of the tumour on the lateral wall. It may extend into the nasopharynx and block the posterior choana. In Type 2 the signs of sinus disease are also present. **Eye signs**—Proptosis alone was present in fourteen cases. It was found associated with optic neuritis and epiphora in thirty-four cases (twenty of these were of Type 2). Optic neuritis is caused either by stretching of the optic nerve on the floor of the orbit or by pressure on the nerve in the optic foramen. **Neurological signs**—Frontal headache and frontal pain suggest involvement of the fifth nerve, especially the supraorbital and infraorbital branches. This headache has been relieved by freeing the nerve from the surrounding bony tissue. Hemianaesthesia of the face and tic doloureux have been observed. The seventh and eighth nerves were not affected in the cases reviewed. ** Mouth signs**—The gums are thickened and oedematous and the teeth show disorderly growth, especially in Type 2. Swelling of the palatal process of the maxilla is more commonly found in Paget’s disease, but was observed in two cases of true leontiasis.

**RADIOGRAPHIC FEATURES**

**Type 1**—The abnormality may take three forms. 1) There is a localised sclerosis beginning in the maxilla and spreading over the frontal bone, and sometimes to the cranium and lower jaw. This is the classical picture. 2) The sclerosis is not complete but is etched out by small translucent patches and areas of normal bone (as in Type 2). This is seen in Frangenheim’s cases and also in my Case 4 (Fig. 9). 3) Sclerosis of the face is associated with a generalised sclerosis affecting other parts of the skeleton, as in Albers-Schönberg’s disease. The sclerosis may be periosteal or endosteal. The antra are dull in Type 1 and often obliterated by the fibro-osseous mass. The sclerosis may affect the cervical vertebrae, but the inner table of the skull is not affected. 

**Type 2**—The following changes may be observed. 1) The facial bones show the typical trabeculated and cystic appearance of a localised osteitis fibrosa. The cortex may be seen as a paper-thin shell (Fig. 2). 2) The base of the skull shows sclerosis, a characteristic finding. 3) The calvarium is thickened and may present an appearance similar to that of Paget’s disease except that the inner table is intact. 4) Osteoporosis circumscripta cranii occurs frequently. It occurs at least as often as it does in Paget’s disease. It is said that 20 per cent of patients with osteoporosis circumscripta cranii show leontiasis or tumours of the maxilla. There were eight cases in this review (six in Type 2 and two in Type 1 cases) (Fig. 9).

**DIAGNOSIS**

The clinical history of proptosis, nasal stenosis and swelling of the face starting in the first decade of life will suggest the diagnosis. The other important investigations are the radiographic examination and biopsy. The whole skeleton should be radiographed. **Biopsy** is important in excluding such conditions as osteoclastoma and osteitis deformans. It is less useful in separating the various types of fibrous dysplasia.

**DIFFERENTIAL DIAGNOSIS**

Symptomatic leontiasis ossea differs from the true variety in etiology, clinical features and pathology. The conditions listed below are those that are most frequently mistaken for leontiasis ossea. Some, such as congenital gigantism of the face, are easy to diagnose. Others, such as congenital syphilitic osteoperiostitis of the face, are almost identical with true leontiasis ossea.
Paget's disease—In the facial region Paget's disease has distinctive clinical features. There is an insidious onset of facial disfigurement, facial pain and deafness arising in the fourth or fifth decades. There is never any interference with vision and the maxilla is affected only in its prealveolar part. The other bone commonly affected is the zygoma. Of eight patients with Paget's disease of the face in this review only one had affection of the lower jaw alone; in all the others the upper jaw was affected. Four had deafness, three suffered from headache, one had poor vision and two had no symptoms beyond facial disfigurement. Only one had nasal stenosis, and proptosis was not observed. The radiographic appearances and histology differ from those of leontiasis ossea.

Partial congenital gigantism—Five cases in the series were examples of partial congenital gigantism, but were listed as leontiasis ossea (those of Kiwul 1890, Clément et al. 1943, Starr 1894, Porter 1904 and Barwell 1881). Partial congenital gigantism of the face or hemihypertrophia faciei et collis (Hamann 1951) is a hypertrophy of all the tissues (including bone) of the face, neck, mouth, tongue and oropharynx, confined strictly to one side. The lips and teeth are most commonly enlarged. It is associated with hyperaemia, pigmentation and marked thickening of the skin. The only atypical case was that of Starr, in which the whole head, whole face and whole neck were enlarged.

Congenital anomalies of the skull—Anomalies of skull development may bear a striking resemblance to leontiasis ossea. The most important is brachycephalus, in which the head is broad, short and rounded. There are three types of this abnormality: 1) oxycephalus (turriccephalus, tower or steeple head); 2) plagiocephalus (asymmetrical skull); and 3) acrocephalus (high projecting forehead). In oxycephalus there is early closure of all sutures, especially the sagittal and coronal. The skull grows upwards into a point. There are exophthalmos, prominent zygomatic bones, enlarged temporal fossae and raised intracranial pressure. Mentality is usually normal. Other defects, such as syndactylism, occur. This is the commonest congenital anomaly of the skull. Plagiocephalus is essentially unilateral oxycephaly. The skull grows upwards, but only on one side. Acrocephalus is believed to be due to premature fusion of the sagittal sutures near the bregma. The frontal bone bulges upwards and forwards. This type is very often associated with syndactylism, a combination known as Apert's syndrome. In all of these three conditions there may be a swelling in the frontal, temporal or zygomatic region, proptosis or exophthalmos, headaches, fits and, frequently, blindness. The conjunction of swollen face, proptosis and disturbance of vision gives the false picture of leontiasis ossea.

In this series there were seven cases of congenital skull anomaly. Two were due to plagiocephaly, three to oxycephaly and two had insufficient evidence on which to classify them.

Other congenital affections, which may be associated with these main defects and give confusion, include Crouzon's disease (cranio-facial dysostosis) in which there are a prominent frontal bone, hooked nose, heavy lower lip, prognathism and high arched palate; Dejerzyński's syndrome (periostitis hyperplastica familiaris), a combination of acrocephaly, syndactylism and a massive skeleton in which there are convolutional atrophy of the brain, a large sella turcica and rudimentary or absent sinuses; hypertelorism (Greig 1924), in which the width between the eyes is increased and there is bulging of the frontal lobes of the brain and protrusion of the frontal bones (Fig. 2); gargoylism (Hurler 1919), in which there may be frontal bossing and facial concavity, with some sclerosis of the bones; and cephalonia, a rare condition in which there is an increase in the brain contents and consequent macrocephaly.

Syphilitic osteoperiostitis—Eight cases of syphilitic osteoperiostitis in this series were listed as leontiasis ossea. Seven were due to latent congenital syphilis and one to acquired syphilis. In half the cases the swellings were paranasal and arose insidiously at about the age of seven and were associated with other syphilitic stigmata. These cases bore a striking resemblance to goundou. But the others were not typical. Ilg's (1821) case showed enormous

THE JOURNAL OF BONE AND JOINT SURGERY
enlargement of the whole face; Pulet-Laherre's (1887) case an opaque sclerosis of the face and cranium. Lapeyre's (1937) case showed the normal distribution and radiographic appearance of true leontiasis of Type 1. It is probable that syphilis in the latter case was coincidental.

The commonest symptom of syphilitic osteoperiostitis was nasal stenosis. Other symptoms were headache, facial neuralgia, dimness of vision and ataxia. Differential diagnosis may be difficult, for almost the only points of distinction from true leontiasis ossea are the syphilitic stigmata, the nasal attachment of the hyperostoses and the usual family history of miscarriages. The paranasal type is very similar to Frangenheim's bilateral familial maxillary hyperostoses. Study of the records, however, shows that there was a vigorous search for syphilitic signs in all Frangenheim's cases without any being found.

**Pituitary dysfunction**—There were six cases of pituitary dysfunction in the series. Two did not even show symptomatic leontiasis ossea. Of the other four, three were due to gigantism and one to acromegaly (the cases of Bassoe 1905, Buhl 1878, Saucerotte 1801 and Sattler 1900). All showed classical hyperpituitarism. In addition they all developed hyperostoses of the maxilla, and all subsequently developed raised intracranial pressure. Acromegaly should never be confused with leontiasis ossea, because of the thick lips and tongue and the marked prognathism. While it is possible that there may be a pituitary factor in leontiasis, acromegaly and gigantism are clinically quite distinct from the true leontiasis.

**Primary bone tumours of the maxilla and nasopharynx**—Three malignant tumours in the series have been named as leontiasis ossea. They were a sarcoma of the maxilla, a carcinoma of the maxilla and a nasopharyngeal fibrosarcoma. The symptoms and signs are sufficiently distinct to need no further differentiation. Benign tumours to be borne in mind include the following: 1) Ivory osteoma, which may be unilateral or bilateral. An osteoma of the frontal sinus may cause proptosis and nasal stenosis. 2) Osteoclastomata, which may easily be distinguished by the radiographic findings and biopsy. 3) Adamantinoma, which has a complex but characteristic pathology. It is found usually in the lower jaw. 4) Ossifying fibroma (fibro-osteoma or fibro-osteoid osteoma). The maxilla is commonly affected, with consequent proptosis. Its pathology is under discussion.

**Other conditions**—Occasionally confusion may be caused by the following: 1) Dyschondroplasia. I found one case in the series and that case showed none of the classical signs of leontiasis ossea. Typically in this disease the face is concave. 2) Meningiomas. There were two possible cases in the series. Many authors refer to the "hemicraniosis of Brissaud and Lerebouliet" as a cause of true leontiasis ossea. In referring to Brissaud's original paper I found the phrase "une double lesion ossée et meningeée." In fact the cases were meningiomas with reactionary bone formation in the fronto-parietal region. The main clinical features are swelling of the forehead or cranium, with headache, vomiting and exophthalmos. The lesion starts first on the inner table, as opposed to the diploe in fibrous dysplasia. 3) Hyperostosis frontalis interna. This is a thickening of the inner table of the frontal bone occurring in women of thirty to forty-five. It may be associated with the Stewart-Morel syndrome, in which two of the common symptoms are dimness of vision and convulsions. 4) Secondary malignant deposits. 5) Erythroblastic anaemia, in which there may be thickening of the frontal bone from proliferation of the diploe. 6) Multiple neurofibromatosis (von Recklinghausen). Beneath the diseased skin, lesions of the bone are often found. They may produce kyphoscoliosis or a congenital dysplasia of one or more bones leading to local gigantism. Cyst-like areas are found in the skeleton. This disease could show many of the features of congenital facial hemi-hypertrophy. In one, gigantism follows the course of nerves, in the other the swelling goes up to the midline. In both there may be hypertrophy of the cervical vertebrae, and pigmentation. Two cases in this series of leontiasis were found to be examples of neurofibroma. In one there was a neurinoma of
the optic nerve and in the other there was generalised neurofibromatosis. 7) Myelosclerosis. A condition of bony thickening and sclerosis is found associated with anaemia, aleukaemic leukaemia and leukaemia. 8) Lipoid granulomatosis. The Hand-Schüller-Christian variety shows clear-cut defects of the skull together with exophthalmos and diabetes insipidus.

PROGNOSIS

The prognosis in leontiasis ossea is better than was thought. In one-third of the patients facial disfigurement is the only symptom. Visual lesions are serious but they occur in only 20 per cent of cases. A patient who has no eye signs at twenty years of age will never develop optic neuritis. It seems probable that the bad prognosis which has been suggested by some is due to the associated diseases that have been listed wrongly as leontiasis. In this series the cases of pituitary adenoma, craniostenosis and syphilitic osteoperoiostitis had a grave prognosis both for vision and life.

Out of ninety-seven patients with true leontiasis ossea thirty-one had no symptoms or signs beyond swelling of the face; twenty-three had nasal obstruction, epiphora or dacrocystitis; nineteen had signs of local pressure on the eye or optic nerve; seven had headache or neuralgia; and eight had symptoms and signs of raised intracranial pressure. There were only four deaths. The incidence of complications was 12 per cent, but they were usually of the mild variety. Perusal of the case histories shows that operation improves the prognosis for vision in nearly all cases and especially if the surgical intervention takes place soon after the onset of symptoms.

TREATMENT

In view of the good prognosis, prophylactic removal of the mass is unjustified. The treatment is mainly symptomatic. The most important step is to check and relieve the eye signs by decompression of the optic nerve. Operations such as Kronlein’s lateral orbitotomy, transfrontal orbitotomy or sub-zygomatic decompression may be necessary according to the position of the growth. Nasal stenosis may require treatment by resection of the septum and chiselling down the floor of the nose. Associated sepsis, in the form of antral empyema, ethmoiditis, dental infection, dacrocystitis or gingivitis may need appropriate treatment. Neuralgia of the fifth nerve, with headache, may require operation. This may be a question of dissecting the nerve out from its ensheathing bone as in Moore’s case. Or it may require injection or section of the ganglia of the nerve or one of its branches.

Probably the commonest surgical treatment in the proved Type 2 cases will be to restore a normal facial contour by plastic operations.

X-ray therapy—This may be used alone or in conjunction with surgery. It has been proved to have a beneficil effect in all cases. There is regression both in the size of the bony tumours and in the symptoms. Dosage recommended is 2,000-3,000r.

CASE REPORTS

Case 1—Girl aged sixteen years. History of gradual onset of swelling of right side of face at age of seven years, after a blow on the cheek. The swelling gradually enlarged and spread to the other side of the face and both lower jaws (Fig. 1). No further increase in size during last eighteen months. Parents dead, but photographs of father and mother showed no facial disfigurement. Child complained of nasal congestion and catarrh. No visual symptoms. On examination, the thickening was found to involve the maxillae, mandible, zygomas (especially right), ethmoids, sphenoid, and base of skull. The calvarium was unaffected. There was bilateral proptosis. There was some limitation of airway in both sides of the nose, from hypertrophic mucosa. Visual activity was 6/5 in each eye; the fundi were healthy; there was slight restriction of both visual fields. Examination of the central nervous system revealed no abnormality. Investigations—Haemoglobin 81 per cent. Red blood cells normal. White cells 10,350 (76 per cent polymorphs). Plasma phosphate 3-1 milligrams per cent; calcium 12 milligrams per cent; cholesterol 115 milligrams and 174 milligrams per cent; alkaline phosphatase 8-3 units. Urinary calcium—Excretion, twenty-four hours, 875 cubic centimetres; inorganic phosphate 1-2 grammes (normal
FIG. 1
Case 1. Figure 1—Multilocular cystic disease of the jaws, or "cherubism," causing leontiasis ossea (Type 2). Note the gross enlargement of the right mandible, right maxilla and right zygoma. There is also proptosis and some swelling of the jaws on the left side. Figure 2—Lateral radiograph of skull, showing the cystic condition of both upper and lower jaws, the sclerosis of the basi-sphenoid and the normal cranium. There is gross malalignment of the teeth.

FIG. 3
Case 1—Photomicrograph of section from maxilla, showing giant cells in a mass of fibroblastic tissue.
1–5 grammes per day); calcium 177 milligrams (normal 10–300 milligrams). *Sternal puncture*—Slight eosinophilia but otherwise normal marrow. *Lumbar puncture*—Normal findings.

**Radiographic examination**—There was a generalised cystic appearance of the facial bones and mandible (Fig. 2). Some trabeculation was present, but it was not of the regular pattern found in osteoclastoma. The cystic changes involved the ethmoidal and sphenoidal air sinuses. There was some sclerosis of the base of the skull, but the calvarium was unaffected. The teeth were ill-developed and malaligned, lying at all angles in the jaws. Some of the molar teeth were unerupted. The antra were opaque. The rest of the skeleton was normal, except for slight notching of the medial aspect of both humeral epiphyses, said to be characteristic of leukaemia (Golding 1950).

**Biopsy**—Material was taken from the right maxillary swelling. *Gross findings*—The cortical bone was paper-thin. The medulla was increased, and filled with a soft, spongy mass. The bone was bluish on its surface, and cut easily. It could be dented by pressure, with a sensation of “egg-shell” crackling. The periosteum as such was not identifiable; the whole subcutaneous area was thickened and fibrotic. *Histology*—The main mass consisted of fibroblasts in an expanded medulla, with scattered giant cells of the osteoclast type (Fig. 3). There were no giant cells in the subcortical fibrous zone. There was no suggestion of osteoid tissue; no lipid and no haemosiderin.

**Treatment**—At the time of biopsy the right maxillary antrum was opened and found to be normal except for the presence of an unerupted canine tooth. The left antrum was opened later and found clear. The disfiguring facial mass was excised by a plastic operation, undertaken in two stages.

**Comment**—The radiographic appearances were striking. They did not resemble those of focal osteitis fibrosa, von Recklinghausen’s disease of bone, osteoclastoma, or odontogenic cyst. But they were like those published by Jones *et al.* (1950) in their cases of familial multilocular cystic disease of the jaws (“cherubism”). Histologically, the profusion of giant cells was unlike the usual findings in fibrous dysplasia or focal osteitis fibrosa, although Thoma (1941) believed that the presence of giant cells was not inconsistent with fibrous dysplasia. Like the radiographic findings, the histological appearances were those of Jones’s cases of cherubism. There was no evidence in this case, however, of a familial incidence. This case is classified as true leontiasis ossea, Type 2, caused by multilocular cystic disease of the jaws.

**Case 2**—Man aged fifty-seven years. Complained of increasing swelling of face which began fifteen years before—about a year after extensive removal of teeth. Had also noticed general enlargement of head. Bilateral deafness and tinnitus for six years. No headache or facial pain.

On examination, there was marked swelling of the upper jaw, especially in the pre-alveolar region (Fig. 4). The lips were protruded by the swelling, but there was no proptosis. Examination of the ears showed normal tympanic membranes. Bone conduction much reduced, especially on the right. Negative rinné test both sides. Nose—There was some hypertrophy of the mucous membrane on the left side, but no nasal stenosis. The eyes were normal.

**Radiographic examination** showed typical appearance of Paget’s disease in the pelvis, and in the maxilla and calvarium (Fig. 5). There was an osteoma of the right frontal sinus and opacity of the left antrum.

**Investigations**—Serum calcium 9·8 milligrams per cent; inorganic phosphorous 2·6 milligrams per cent; alkaline phosphatase 15·3 units.

**Treatment**—Left intranasal antrostomy was undertaken for relief of a chronic empyema of the left antrum. No other treatment was considered necessary.

**Comment**—This case is classified as false leontiasis ossea due to Paget’s disease involving the maxilla. Distinctive features, not present in true leontiasis, were the pre-alveolar swelling and perceptive deafness, and the absence of nasal stenosis or proptosis.

**Case 3**—Girl aged seven years. Insidious onset of right facial swelling and right proptosis for three years. The swelling had increased gradually. Tendency to right epiphora. No neurological or visual symptoms.

On examination there was swelling of the right zygoma and right temporal region, with proptosis (Fig. 6). The eyes were widely spaced, the right being lower than the left. The ophthalmologist reported hypertelorism in a congenitally defective skull; vision 6/18 each eye; alternating divergent and paralytic squint; media clear; fundi normal.

**Radiographic examination**—The lesion affected the skull, right zygoma, right temporal bone, parts of right parietal and right frontal zone. The skull showed the “beaten silver” appearance of
Case 2—Paget’s disease of skull and face causing false leontiasis ossea.

Fig. 4

Fig. 5
Case 2—Radiograph showing typical Paget’s appearance of the skull and face.
craniosynostosis, with a bony thickening in the region of the right zygoma (Fig. 7). Tendency to plagioccephaly.

Treatment—Expectant treatment has been planned.

Comment—This case is classified as false leontiasis ossea due to craniosynostosis. The bony hypertrophy was marked. This, with the proptosis, rendered confusion with true leontiasis easy. The cause of the bony hypertrophy was presumably connected with alteration of the bony mechanics of the skull; the cranial contents were protruded in those areas in which there was no premature synostosis.

Case 4. Man aged fifty-nine years. At almost the age of twenty years he first noticed a swelling of the left side of the forehead, which slowly increased in size for ten years. He attributed the onset of the swelling to a blow on the head. The swelling was symptomless; it was observed when he reported for treatment of osteomyelitis of the left maxilla. The osteomyelitis responded to penicillin. There was a history of old bilateral otitis media.

On examination, the main swelling affected the left frontal area, which was thickened as far back as the left parietal suture (Fig. 8). There were other exostoses in the left parietal suture and left occiput, but these were separate from the main mass. Both maxillae and both zygomas were enlarged. Ears—There was deafness due to old bilateral supplicative otitis media. Eyes—The visual field on the nasal side of the left eye was restricted.

Radiographic examination showed that both maxillae and the left frontal, parietal and occipital bones were the seat of fibrous dysplasia. There was sclerosis of the base of the skull, the face, and the frontal region especially (Fig. 9). Farther back there were cystic areas typical of fibrous dysplasia and localised areas of osteoporosis circumscripcta cranii. The rest of the skeleton was normal.

Investigations—Serum calcium 10·2 milligrams per cent; phosphate 4·2 milligrams per cent; alkaline phosphatase 20·2 units. Urinary calcium excretion was normal. An osteomyelitic cavity in the left maxilla was excavated. The removed bone showed only the changes of osteomyelitis. Biopsy of the main mass was not undertaken.

Comment—The appearance of the calvarium suggested true leontiasis ossea of Type 2 (fibrous dysplasia). Nevertheless there was so much sclerosis anteriorly that the case was classified as true leontiasis Type 1, associated with osteoporosis circumscripcta cranii. Strictly,
Case 4—Truc leontiasis ossea (Type 1). Note the marked enlargement of the left maxilla and left frontal region.

Fig. 8

Case 4—Radiograph showing the fibro-dysplastic areas in the cranium, and sclerotic bony hypertrophy of the base of the skull, the frontal bone and the maxillae. In the frontal bone the mottled appearance characteristic of fibro-dysplasia is clearly seen. (See text for discussion on diagnosis.)

Fig. 9
it is a mixed type. The development of osteomyelitis suggests that the fibrous bone is more susceptible to infection than normal bone. It has been noted that when fibrous dysplasia begins relatively late in life, as in this case, the tendency is for the bone to become more sclerotic than usual, and for the symptoms to be slight.

SUMMARY AND CONCLUSIONS

1. One hundred and forty-nine cases of leontiasis ossea reported in the literature have been reviewed.
2. The clinical, radiographic and pathological features of the condition are discussed.
3. Four additional personal cases are reported in detail.
4. A new classification of leontiasis ossea is suggested, by which the condition is divided into true leontiasis and false (or symptomatic) leontiasis. True leontiasis is a clinical syndrome caused by two distinct types of disease, whose pathology, however, is related. False leontiasis gives a superficially similar picture but on detailed examination is found to be distinct; it may be caused by a variety of different conditions.
5. Classified in this way, ninety-seven of the total of 153 cases considered were classified as true leontiasis (forty-five Type 1 and fifty-two Type 2); forty were classified as false leontiasis; and sixteen did not fall into either category.

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LEONTIASIS OSSEA

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