VERTEBRAL HYDATIDOSIS AND PARAPLEgia

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We report the management of two children and 11 adults with paraplegia secondary to vertebral hydatidosis. Destruction of pedicles, posterior vertebral elements and discs as well as the vertebral bodies was common and all six patients with thoracic disease had involvement of adjacent ribs.

The 13 patients had a total of 42 major surgical procedures; two patients died from postoperative complications and four from complications of the disease and paraplegia. All eight patients initially treated by laminectomy or anterior decompression alone relapsed within two years and seven required further surgery. Circumferential decompression and grafting gave the best results, six of nine patients being in remission an average of three years and six months later.

The prognosis for such patients is poor; remission is the aim, rather than cure. Anthelminthic drugs may improve the prognosis, but radical surgery is likely to remain the keystone of treatment in the foreseeable future.

The incidence of human infestation with *Echinococcus granulosus* is increasing, and the parasite is appearing in areas of the world previously free of it (Williams, López, Adaros and Trejos 1971; Matossian, Rickard and Smyth 1977). We report the treatment and results of 13 patients with paraplegia due to vertebral hydatidosis.

**PATIENTS AND FINDINGS**

From 1965 to 1986 13 patients with vertebral hydatidosis and paraplegia were admitted to the Mohamed Kassab National Orthopaedic Institute at Kassar Said, Tunisia. There were seven male and six female patients aged between 10 and 65 years (average age 33). The average delays were: onset of symptoms to first medical consultation, 15 months (range 2 to 72); first symptom to first neurological sign, 22 months (range 0 to 68); and onset of paraparesis or paraplegia to the first decompressive surgery, six months (range 2 to 14).

The presenting symptom was localised back pain in seven patients, radicular leg pain without back pain in

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Fig. 1

Radiograph of the lumbar spine of a 29-year-old paraplegic man two years after partial laminectomy at three levels. There is lateral destruction of the body of the fourth lumbar vertebra and partial destruction of the proximal intervertebral disc and the right transverse process. The distal intervertebral disc is completely destroyed.
one, while two patients had both. The other three patients presented with spastic paraplegia, flaccid paraplegia and a cauda equina syndrome respectively. Four patients had an initial erroneous diagnosis of Pott's disease and had been treated with antituberculous drugs.

When first seen at the Institute three patients with dorsal and two with lumbar disease had flaccid paraplegia, three with dorsal disease had spastic paraplegia, and five with lumbar disease had paraparesis. Seven patients had developed intra-abdominal or pulmonary cysts, and the immuno-electrophoresis test was positive in six of the seven patients tested. In all 13 patients the diagnosis was confirmed at surgery.

The parasite had affected the dorsal spine in six patients, the lumbar in five and the lumbosacral junction in two. One vertebra was involved in two patients, two vertebrae in eight and three vertebrae in three patients. In all cases there were osteolytic lesions in the vertebral bodies and in 12 the adjacent discs were destroyed (Fig. 1). Seven patients showed an absent pedicle and five had destruction of the posterior arch. In all six patients with thoracic disease the adjacent ribs were also affected (Fig. 2a). Ten patients had a short angular kyphosis or kyphoscoliosis averaging 62° (maximum 105°) (Figs 1, 2a, and 3). Three patients had a thoracic paravertebral abscess, one had a psoas abscess and one a pelvic abscess. Nine patients had a pre-operative myelogram (Fig. 3). In one of these the dye had outlined an abscess cavity which communicated with the subarachnoid space.

TREATMENT AND RESULTS

Six patients had had 11 posterior decompression operations before referral to us, carried out for spastic (2) or for flaccid (4) paraplegia. Five of these patients had had some temporary improvement in neurological status and
one was unchanged, but all six patients had relapsed three months to two years after the primary operation and before referral. All the recurrences were due to the incomplete removal of diseased tissue.

At our Institution, one patient had an anterior resection of the vertebral body and abscess mass 18 months after laminectomy, but died postoperatively in renal failure due to a mismatched blood transfusion. One patient had a second laminectomy for two draining sinuses and was then lost to follow-up.

Four patients were treated by double operations, that is the anterior approach followed two to six weeks later by the posterior approach, with excision of as much of the diseased bone and surrounding tissue as possible and grafting with tibia or fibula anteriorly and iliac crest bone posteriorly. One of these patients developed a pseudarthrosis anteriorly and was graft ed again six months later, and one patient was stabilised posteriorly with Roy-Camille plates. Three of the four patients had partial neurological improvement and were still in remission at 6, 18 and 60 months after operation. One patient, who had had four previous laminectomies had complete neurological recovery, but relapsed with paraplegia 13 months later and died.

Seven patients had had no previous surgery. We treated one by laminectomy alone, with no improvement and death three years later. One patient had anterior decompression and bone grafting but his neurological status deteriorated with cauda equina compression by the graft which was not relieved by re-operation. He also died shortly after; both deaths were from complications secondary to paraplegia.

The other five primary patients were treated by a double approach, two having posterior Harrington instrumentation for instability (Fig. 2). One patient died as a result of massive haemorrhage from the common iliac artery which had been eroded by the cyst. One other patient with disease so extensive that complete removal was impossible was not improved and died 28 months later. The remaining three patients were initially improved: one still had complete recovery at 32 months follow-up, but the other two relapsed within 18 months and were treated again by the anterior approach. One of these was still in remission 54 months later, the other had a wound infection and meningitis, then required a thoracotomy for abscess, but his neurological status improved markedly, and he was still in remission at 78 months.

**DISCUSSION**

Vertebral hydatidosis is said to be a relatively silent, slowly progressive disease with a latent period of many years (Dew 1928; Mills 1956; Alivisatos and Spiliotis 1957). We agree with Allred and Nisbett (1964), Apt et al (1976) and Saidi (1976) that pain is an important presenting symptom. Vertebral hydatidosis is rare in children. Gharbi et al (1977) reported one case in a 13-year-old, and Slim et al (1971) found no cases among 34 children with hydatid disease of other organs. Two of our patients were aged 10 and 13 years at diagnosis, and both showed rapid progression.

There are few characteristic radiological features since the parasite tends to spread in all directions (Dévé 1928; Bellini 1946; Acquaviva and Tamic 1964; Carta, Perria and Davini 1974). Bone is destroyed by compression without reactive new bone formation (Gangolphe 1886; Schroeder and Medoc 1952), the cortex being eroded or destroyed with the subsequent formation of an 'abscess' in the surrounding soft tissues. Adjacent vertebrae may be involved by extension beneath the longitudinal ligament or through the disc space. The disc is reported to be relatively resistant to invasion (Bellini 1946; Froment et al 1984), and Murray and Haddad (1959) stated that a soft tissue shadow next to destroyed vertebrae with intact discs should suggest the diagnosis of hydatidosis. However, 12 of our patients had some destruction of adjacent discs.

Involvement of an adjacent rib is peculiar to hydatidosis (Bellini 1946; Dévé 1948; Froment et al 1984). Griel and Dévé (1929) noted that 37% of 129 thoracic cases had an adjacent rib affected, and Froment et al (1984) reported this double pathology in 50%. In our series all the patients with thoracic disease had rib involvement.

Paraplegia is the most serious complication of vertebral hydatidosis; cysts invade the spinal canal and cause direct compression or ischaemic changes in the spinal cord or the cauda equina (Woodland 1949; Robinson 1959; Booz 1972; Braithwaite and Lees 1981; Froment et al 1984). Battaieb et al (1978) stated that radicular pain precedes paraparesis by a few weeks and there is rapid progress to paraplegia in the thoracic spine, but slow progress in the lumbar spine. In contrast Saidi (1976) reported slow progression of neurological compression. Three of our patients with thoracic disease developed sudden paraplegia.

Schroeder and Medoc (1952) consider that spinal cord damage is often reversible, but Turtas, Viale and Pau (1980) disagreed and advised early operative decompression before irreversible changes occur. In our series, in which no patient had early operation, eight had neurological improvement after the first decompression and four did not. One died before any change could be assessed. This suggests that surgery is worthwhile however long the cord or cauda equina has been compressed.

In hydatid disease, standard laboratory tests such as the ESR and eosinophil counts have little specific diagnostic value and even the intradermal Casoni test is uncertain. Several specific tests have been described (Capron et al 1970; Pinon and Dropsy 1976; Chemtai, Bowry and Ahmed 1981); we used the immuno-electrophoresis test and this was positive in 87% of the patients tested.

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The differential diagnosis of vertebral hydatidosis includes Pott’s disease and primary or secondary tumour (Robinson 1959). In Pott’s disease there is always some bony reaction to the destructive lesion and usually early narrowing or destruction of the adjacent disc, but the infection very rarely involves a pedicle, the posterior arch or a contiguous rib (Moula et al. 1981).

The prognosis of vertebral hydatid disease is poor, especially when neurological symptoms and signs are present. In Britain the average length of survival after the onset of symptoms was five years, the average age at death being 41 (Mills 1956). Treatment is essentially surgical, traditionally by decompressive laminectomy (Dew 1928; Alldred and Nisbet 1964; Bettiaieb et al. 1978), but this is rarely curative because it does not allow excision of the diseased vertebral bodies and adjacent tissues. Most patients reported in the literature have had several operations for recurrence (Alivisatos and Spiliotis 1957; Robinson 1959; Apt et al. 1976; Ferrandez et al. 1978; Turtas et al. 1980), and the outlook is so gloomy that Acquaviva and Tamic (1964) took a ‘second look’ in nearly half of their patients and Guedj and Marill (1963) advocated this in all patients.

In our series 13 patients had a total of 42 major surgical procedures. None of the eight patients treated by laminectomy or anterior decompression alone had permanent improvement. All seven survivors required further surgery for recurrence and neurological deterioration within two years of the first operation, and three of these are known to have died.

Vertebral hydatidosis can be considered a locally malignant tumour, the ‘cancer blanc’ of Dévé (1948) (Dew 1928; Alldred and Nisbet 1964; Bettiaieb et al. 1978). Ideally, treatment would be by total excision of all affected tissue with a shell of healthy tissue, but this is unattainable because of the close proximity of vital structures. Palliative resection of the diseased focus and the associated abscess cavities is however possible through a double approach (Del Campo 1950; Alivisatos and Spiliotis 1957; Guedj and Marill 1963; Savini and Capelli 1978). In our series nine patients were treated in this way, four after previous failed surgery. Three of these patients died, but six were still in remission an average of 3 years 6 months after their last operation.

The scleroidal effects of the anthelmintic drugs mebendazole, albendazole and flubendazole for soft tissue infestation by Echinococcus granulosus have been described by Heath and Chevis (1974) and by Morris et al. (1983). The value and safety of these drugs in vertebral hydatidosis is uncertain (Webster 1985; Coulaud 1988), but some authors feel that they may improve the prognosis when combined with surgery (Fiennes and Thomas 1982; Cardona et al. 1983; Porat, Robin and Wertheim 1984; Savini et al. 1984; Fournier et al. 1985; Ocete et al. 1986). We did not use these drugs during the period of this investigation.

Vertebral hydatidosis with paraplegia is a lethal disease, reported mortality rates varying from 14% to 58% for a mixture of patients with and without paraplegia (Schroeder and Medoc 1952; Mills 1956; Acquaviva and Tamic 1964; Alldred and Nisbet 1964; Ferrandez et al. 1978; Turtas et al. 1980). The known mortality rate in our series of 13 patients is 46% and this may increase with a longer follow-up.

Prevention of the disease by eradication of the parasite from the pool of primary hosts, as was done in Iceland and New Zealand, is the only way to avoid such suffering for so many people (Dungal 1957; Gemmell 1958). However, the parasite grows slowly and surgery can often ameliorate the symptoms and prolong life for many years (Dew 1928; Schroeder and Medoc 1952; Alivisatos and Spiliotis 1957). We hope that the combination of anterior and posterior surgical approaches with anthelmintic drugs will markedly improve survival rates.

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REFERENCES


