Ankylosing neurogenic myositis ossificans of the hip
AN ENHANCED VOLUMETRIC CT STUDY


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Neurogenic myositis ossificans is a disabling condition affecting the large joints of patients with severe post-traumatic impairment of the central nervous system. It can result in ankylosis of the joint and vascular or neural compression. Surgery may be hazardous with potential haemorrhage, neurovascular injury, iatrogenic fracture and osteochondral injury. We undertook pre-operative volumetric CT assessment of 45 ankylosed hips with neurogenic myositis ossificans which required surgery. Helical CT with intravenous contrast, combined with two- and three-dimensional surface reconstructions, was the only pre-operative imaging procedure. This gave good differentiation of the heterotopic bone from the adjacent vessels. We established that early surgery, within 24 months of injury, was neither complicated by peri-operative fracture nor by the early recurrence of neurogenic myositis ossificans. Surgical delay was associated with a loss of joint space and a greater degree of bone demineralisation. Enhanced volumetric CT is an excellent method for the pre-operative assessment of neurogenic myositis ossificans and correlates well with the operative findings.

Neurogenic myositis ossificans is the formation of heterotopic bone in the periarticular soft tissues of patients who have sustained a severe injury to the brain or spinal cord. It normally occurs in the first weeks or months after the injury. Although neurogenic myositis ossificans may develop in any joint, it usually involves the large joints, often the hips and, less commonly, the knees, elbows and shoulders.

It is a disabling complication which may cause severe limitation of joint mobility or ankylosis, and vascular or neural compromise. Surgical removal of mature heterotopic bone is required in order to regain a functional range of movement although the surgeon must take care to avoid fractures and damage to adjacent neurovascular structures. Scintigraphy, arteriography and phlebography are normally performed before such surgery. In our study, however, pre-operative planning was undertaken by volumetric CT only.

Patients and Methods
Between 1995 and 2002, we performed a prospective radiological and clinical study of 29 adult patients (45 hips). There were seven women, and 22 men with a mean age of 45.5 years (19 to 69).

Twelve patients had suffered cerebral trauma, seven trauma to the spinal cord, five vascular disease and five complications as a result of a lengthy stay in intensive care. No patient had a history of previous dislocation or hip replacement.

Surgery for neurogenic myositis ossificans was performed by the same orthopaedic surgeon (PD) at a mean of 44.5 months (7 to 185) after the initial trauma. The mean follow-up period was 45.5 months (8 to 84).

Technique of volumetric CT. A pre-defined CT technique was used. Because of severe deformities, the first step was to position the patient. Deformities were corrected as much as possible by various methods such as the use of cushions. Intravenous contrast medium was then administered. After placing tourniquets around the proximal part of the affected limb, a distal vein was catheterised followed by an initial bolus injection of 60 ml of an iodinated solution (meglumine ioxitalamate) of 300 mg/ml. One minute after the end of the first bolus, an iodinated solution (sodium ioxitalamate) of 120 mg/ml was power-injected at an injection rate of 3 ml/s. This technique allowed good differentiation of bone, arterial and venous densities.

Volumetric CT (Philips CT Twin, Eindhoven, Holland) was performed on all patients during the power injection. Axial reconstructions, 3.2 mm in thickness every 1.6 mm, were obtained, as well as two-dimensional multiplanar recon-
structions in the coronal and sagittal planes. Three-dimen-
sional surface reconstructions were then performed based
upon density colour encoding. This technique allowed the
bones to be clearly visible in white, the veins in blue and the
arteries in red. No patient underwent a bone scan before
surgery in order to assess the maturity of the ossification.

CT assessment of articular changes. Pre-operative volumetric
CT was performed one month or less before surgery. This
determined the location and sites of attachment of neuro-
genic myositis ossificans to the bones, as well as the volume,
morphology, borders, and any fragmentation and/or pseu-
darthrosis. Its relationship to blood vessels and nerves
could also be studied.

Our analysis focused on joint changes since ankylosis is
often responsible for joint lesions which can lead to per-
operative difficulties. The CT analysis examined specifi-
cally: 1) location (anterior, posterior or circumferential); 2)
shape (limits, continuity or pseudarthrosis, fragmentation);
3) relationship to the joint capsule (capsular contact or dis-
ruption); 4) bone fragility. The bone density of the femoral
head was compared with that of the ilium, immediately
above the acetabulum. Bone mineralisation was classified
into one of four categories, as follows: normal (M1; Fig. 1);
mild demineralisation (M2; Fig. 2); significant deminerali-
sation with a risk of fracture (M3; Fig. 3); and evanescent
bone with a predictable fracture (M4; Fig. 4). The thickness
of the joint space was also assessed and classified into one
of three categories as follows: normal (S1); narrowing (S2);
and osseous ankylosis (S3; Fig. 5).

A full surgical report was made after each operation,
including a description of the principal site of ossification,
its shape, and its relationship to the joint capsule. Peri-oper-
avtive mobilisation and resection of ossification was some-
times associated with fracture of the hip which was treated
by arthroplasty.

All the patients were reviewed every six months or when
they were referred by their physicians with complications.
Follow-up examinations focused on fracture, infection of
the prosthesis and any recurrent ossification.

Statistical analysis. For statistical comparisons of groups,
Student’s t-test was used. A p value of < 0.05 was regarded
to be significant.

Results

CT analysis. The site of ossification was anterior in 25 hips,
posterior in eight and surrounding the joint in 12. In 29
S3 score illustration. Figure 5a – Axial CT views of a patient with ankylosis of both hips 74 months after an injury to the spinal cord with total paraplegia. The joint space has completely disappeared and there is continuity of bone between the femoral head and the acetabulum. Figure 5b – Frontal and three-dimensional surface CT reconstructions show a total absence of joint space.
hips, the ossification had regular margins but in 16 it had irregular margins and a pseudo-infiltrative appearance. In all patients neurogenic myositis ossificans was continuous, except in one in whom there was clear discontinuity and an obvious pseudarthrosis. A single bony bridge was visible in 24 hips, but in 21 the neurogenic myositis ossificans comprised several osseous fragments.

Apart from two hips, in which there was no contact between neurogenic myositis ossificans and the joint capsule, all showed focal or extensive capsular contact. In two of these capsular disruption was seen.

In terms of fragility, the bone was considered to be M1 in four hips, M2 in 12, M3 in 13 and M4 in 16. The mean time between injury and surgery was 10.75 months for M1 hips, 10.08 months for M2, 35.23 months for M3, and 86.56 months for M4. There was a significant difference in this delay between the combined M1 and M2 groups and the combined M3 and M4 groups (p < 0.001). The width of the joint space was normal in 31 hips, narrow or absent in seven and ankylosed in seven.

Operative findings. We confirmed the CT description for the site, shape and relationship to the joint capsule of neurogenic myositis ossificans by surgical exploration of each hip. In 14 hips, a fracture of the femoral head occurred during, or immediately after, excision of the heterotopic bone. Twelve were treated by an immediate total hip arthroplasty (THA) and two by a delayed arthroplasty after resection of the femoral head and neck. An early post-operative fracture occurred in both hips of one patient with bilateral ossification. Bilateral THAs were performed. In terms of bone fragility all hips which fractured were classified as M4. There was a significant difference in the delay to surgery between fractured and non-fractured hips (p < 0.001). Four of the 14 THAs became infected. There was recurrent ossification in two patients (two hips), in one after early operative treatment of the neurogenic myositis ossificans (13 months). No further surgical treatment was required as there was no functional disability. The other recurrence was seen after late surgical treatment of neurogenic myositis ossificans (60 months). This patient required further surgery 15 months later although a delayed fracture of the hip occurred after this second operation.

Discussion

First described by Dejerine and Ceillier and Dejerine, Ceillier and Dejerine, periarticular ossification arising in patients with severe neurological disorders has been termed heterotopic ossification, para-articular ossification, hyperostosis and myositis ossificans traumatica. This varied terminology has been confusing and has led to these lesions being mistaken for non-traumatic myositis.

Neurogenic myositis ossificans occurs most often in immobile patients with head or spinal-cord injury. It may also be seen as a complication of other protracted diseases of the central nervous system such as inflammatory, toxic, vascular or neoplastic lesions and has been described in bedridden, comatose patients or as a complication of tetanus. Although neurogenic myositis ossificans lesions are seen in large joints, their location and incidence depend upon the underlying disease. Shoulders and elbows are more often affected after head injury, whereas hip and knee more often in the spinally-injured patient.

The incidence of neurogenic myositis ossificans after head injury does not seem to be well established. After injury to the spinal cord, it is said to be between 15% and 20%.

Symptoms may develop within one to ten months but usually occur two to three months after injury.

Many hypotheses about the physiopathology of neurogenic myositis ossificans have been proposed. Some authors have linked its appearance to vasomotor, metabolic and trophic disorders induced by prolonged, total immobility. Others considered that stasis in the paravertebral venous plexus, caused by immobilisation, added to skeletal demineralisation, was the main factor responsible for the precipitation of calcium in the periarticular soft tissues.

A further theory is that the induction of enchondral ossification may result from repeated tendinous or muscular microtrauma caused by forcible, passive mobilisation of the paralysed limb during rehabilitation. This theory has been supported by experimental evidence.

Prophylactic treatment has been described for heterotopic ossification occurring after THA or following the surgical treatment of acetabular fractures. The incidence of heterotopic ossification after surgery for acetabular fracture is significantly reduced by irradiation or anti-inflammatory medication (7% to 38%). Indometacin is as effective as irradiation. Several forms of medical treatment have been tried for early neurogenic myositis ossificans but without significant benefit. Ultrasonography can diagnose these early lesions before ossification becomes evident on plain radiography.

In order to facilitate rehabilitation, surgery for neurogenic myositis ossificans must be performed as early as possible. This is risky since potential complications include injury to demineralised bones or to compressed and entrapped vessels or nerves within the lesions. Pre-operative planning is vital. Helical CT, with multiplanar and three-dimensional reconstructions, is well adapted to this. The special injection technique which we used improved vascular visibility, as well as the differentiation between arteries and veins. Three-dimensional helical CT can also detect thrombophlebitis, which is often associated with neurogenic myositis ossificans. Three-dimensional reconstruction identifies the relationship between the vessels and the ossification. Arteriography and phlebography can therefore be avoided. Precise identification of nerves remains difficult since they are rarely visible on CT when bulky, osseous lesions are present. It may occasionally be possible to see them by identifying a groove containing the nerve in the heterotopic bone mass.
The patients in our study had been referred to us for disabling neurogenic myositis ossificans with hip ankylosis. When possible we operated on these patients early although sometimes the delay to surgery was very long, with a maximum of 185 months.

We found that CT assessment was very useful for determining the degree of bone demineralisation. Assessment of bone demineralisation by osteodensitometry is not appropriate because of the heterotopic ossification within the NMO lesions. CT densitometry is also unsuitable for the hip because there is no phantom available for calibrating the equipment.

We used the iliac bone as our reference for mineralisation since we had noted that its trabecular architecture and mineralisation were largely maintained even in patients after long-term immobilisation.

Although our assessment of hip demineralisation lacked precision, we were still able to correlate it with the surgical findings. Peroperative, or immediately post-operative, fractures only occurred in hips with a mineralisation score of M4. This was a statistically significant relationship. When surgery was performed before 24 months, neither an M4 score nor peroperative fracture was found. This implies that surgery should be performed earlier rather than later.

Narrowing of the joint space was easier for us to determine by CT than by attempting to identify chondral changes by CT arthrography or MRI. Spontaneous arthropathy was only easily demonstrated by the presence of continuous bone trabeculae between the acetabulum and the femoral head. We found a significant association between the delay to surgery and narrowing of the joint space, implying that surgical delay increases the risk of alteration to the joint space which may in turn, impair subsequent function of the hip. All patients who underwent surgery 36 months or more after their initial injury showed narrowing of the joint space. All those who underwent surgery before this time had a normal joint space, even the patient with a demineralisation score of M4.

There was recurrent ossification in only two hips (4.4%) on radiological follow-up. One occurred after early surgery and the other after late surgery. Theoretically, recurrence takes place when surgery has been haemorrhagic, or when maturation of neurogenic myositis ossificans is inadequate, with a persistence of inflammatory areas. Reappearance of neurogenic myositis ossificans is seen within these inflammatory portions. Bone scanning was classically used as an indicator of osseous activity and maturation and surgery was not performed until it became negative. This resulted in delayed surgery and a higher risk of bone trauma. However, there is evidence to support our findings, that surgery should be performed early since demineralisation and joint destruction correlate with a prolonged surgical delay.

Those patients in our study who sustained a peri-operative or immediate post-operative fracture of the hip received an immediate or delayed THA. Unfortunately, 25% of these patients developed septic loosening probably facilitated by recurrent infection of the urinary tract which is so common in paraplegic patients. Our findings thus suggest that early surgery (before 24 months) of ankylosing neurogenic myositis ossificans is not complicated by fracture of the hip. In contrast with some reports, recurrence of ossification does not appear to be more frequent after early surgery. Helical CT with contrast media is the only useful preoperative imaging technique. Isotope bone scanning, arteriography and phlebography are probably now unnecessary.

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References