ILIACUS HAEMATOMA

A Common Complication of Haemophilia

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This paper describes and seeks to explain a clinical syndrome which occurs characteristically, but not exclusively, as a complication of haemophilia. The main features of the syndrome are the sudden onset of severe pain in the groin accompanied by marked flexion deformity at the hip, followed by a mass in the iliac fossa and femoral nerve palsy.

Episodes of this sort have been reported in the literature since the publication of Getting's case by Bulloch and Fildes in 1911. We have found only fourteen published cases since then (Günther 1924, 1935; Seddon 1930; Tallroth 1939; Aggeler and Lucia 1944; Weil 1946; Lyons 1953; Douglas and McAlpine 1956; Hall 1961). The syndrome is common in haemophiliacs: of forty patients studied by Davidson, Epstein, Miller and Taylor (1949) no less than fifteen had suffered at least one such episode. The syndrome has usually been attributed to haemorrhage into the psoas muscle, and Weil (1946) considered this the commonest site of intramuscular haemorrhage in haemophilia.

We have studied twenty-four patients, eleven of whom were admitted to the Nuffield Department of Orthopaedic Surgery for treatment of this syndrome from 1962 to 1965. The other thirteen had been treated elsewhere and were examined by one of us. We believe that the syndrome results from haemorrhage into the iliacus muscle.

ILLUSTRATIVE CASE REPORT

The following case history is in many ways typical. In October 1962 a boy of sixteen who was known to suffer from Christmas disease complained of pain in the left hip. On the day after onset the pain became worse and he was admitted to hospital and given transfusions of plasma. Two days later hypoaesthesia in the distribution of the left femoral nerve and weakness of the quadriceps muscle were noticed, and a mass was felt in the left iliac fossa. He was transferred to the Nuffield Orthopaedic Centre the same day. He had severe pain in the left iliac and lumbar regions and held the left hip flexed to 30 degrees. It could be flexed a further 30 degrees and there was a fairly free range of abduction, adduction and rotation. Cutaneous sensibility was impaired in the area of distribution of the femoral nerve; there was paralysis of the quadriceps muscle, and the patellar tendon reflex was absent. Deep in the left iliac fossa a tender mass was felt extending laterally along the iliac crest. In the past he had suffered numerous haemarthroses and two episodes of haematuria. Both knees and both elbows lacked full movement.

Treatment—He was treated in bed with the left thigh supported in flexion and 500 millilitres of Christmas factor (factor IX) concentrate were given by intravenous infusion. This was of activity equivalent to six litres of normal plasma, and raised his plasma Christmas factor concentration from nothing to 118 per cent of average normal. A second dose was given on the following day. Pain was persistent and severe for the first twenty-four hours, being poorly controlled by large doses of pethidine, after which it slowly diminished. For two weeks he had intermittent discomfort, but the size of the mass in the iliac fossa remained unaltered. A month after the onset he got out of bed and the next day the pain returned and the mass became much larger. Two separate swellings could now be felt; a rounded one filled the cavity of the

inner aspect of the ilium, and a fusiform one was felt medially, extending upwards in the line of the psoas muscle. The combined swellings almost filled the lower left abdomen. The femoral nerve palsy was complete. The patient was put on a Robert Jones frame with the left hip flexed, and further Christmas factor was given. The pain rapidly subsided and in two weeks the mass was much smaller. A month later he was taken off the frame and the joints were mobilised in the swimming pool under cover of Christmas factor and plasma transfusions. At that time the mass had almost disappeared but the femoral nerve palsy was still complete. Fifteen months later the quadriceps femoris muscle had regained full power, but the patellar tendon reflex was absent. Sensory loss was limited to a small area over the patella and no mass was palpable in the abdomen.

THE SYNDROME

The following description of the syndrome is based on observations made on the twenty-four patients we have examined and on fourteen case histories published by other authors. All but one of these patients suffered either from classical haemophilia or Christmas disease, and had suffered previous bleeding. Most had had many haemarthroses, particularly in the knee. One patient, not a sufferer from a bleeding disease, developed an iliacus haematoma complicating anticoagulant therapy with phenindione and we believe that this is the first report of the syndrome occurring other than in one of the bleeding diseases.

Presenting symptom—Pain was always the presenting syndrome. It usually started in the groin and spread to the lumbar region or the thigh. Onset was sudden in some patients and insidious in others, but the intensity steadily increased until it was severe and constant. In none did severe trauma precipitate the haemorrhage, but in several patients the onset of pain followed strenuous use or a minor twist of the limb.

Flexion contracture of the hip—The hip was always held in flexion and usually in lateral rotation. The patients assumed a characteristic posture in bed, lying curled up, often on the affected side, with the hips and knees flexed. Extension of the hip made the pain worse but other movements were present if extension was avoided. This sign distinguishes iliacus haematoma from haemarthrosis or acute suppurative arthritis of the hip.

Femoral nerve palsy—Haemorrhage into the iliacus muscle is usually followed by femoral nerve palsy, but not always (Tallroth 1939, Weil 1946); we have seen one patient in whom an undoubted iliacus haematoma did not cause nerve compression. In many reported cases the nerve lesion was not detected until several days or weeks after the development of the haematoma, but in our experience nerve compression begins with the onset of pain or very soon afterwards. Nerve palsy is limited to the distribution of the femoral nerve and usually becomes complete. The quadriceps femoris muscle is paralysed and the patellar tendon reflex absent. Figure 1 shows the distribution of cutaneous sensory loss in one of our patients and is typical. The area of total anaesthesia varied with the degree of overlap by adjacent nerves but the skin over the patella was always anaesthetic.

Lyons (1953) reported a patient with palsy of the lateral cutaneous nerve of the thigh as well as the femoral nerve. One of us has examined two haemophilic patients who had anaesthesia of the lateral surface of the thigh after a "haemorrhage in the groin" but no other evidence of nerve lesion. We believe that iliacus haematoma is in the great majority of cases accompanied by an isolated femoral nerve palsy, that it occasionally occurs without

FIG. 1

The shaded area on the thigh and leg shows the extent of skin anaesthesia. The shaded area on the abdominal wall is the surface marking of the iliacus haematoma which was palpable above the iliac crest and inguinal ligament.

nerve compression at all, and that rarely the lateral cutaneous nerve of the thigh may be involved.

Mass in the iliac fossa—There was tenderness in the groin and guarding in the iliac fossa in all cases. In some the greatest tenderness was about the mid-inguinal point and slight fullness could be felt there. In others the tenderness was greatest in the iliac fossa next to the iliac crest. It was not possible to feel the mass in patients examined soon after the onset of symptoms as the haematoma takes some time to reach its full size, and "guarding" makes palpation difficult. It always became palpable after one or two days.

FIG. 2

The age at onset of thirty episodes in twenty-four patients. The patient over 65 years was receiving anticoagulant therapy.

The mass arises from the lateral wall of the pelvis and makes the normal concavity of the inner aspect of the wing of the ilium convex. In three of our patients the tumour filled the iliac fossa and a second, discrete, fusiform tumour became palpable medial to the first, with a groove between the two. This second swelling was caused by blood distending the psoas sheath.

Anaemia—Anaemia developed a few days after the onset of symptoms but the blood loss was never enough to cause shock.

Constipation—Constipation, abdominal distension and colic occurred in most of our patients. Bowel sounds were present, but diminished, and two patients needed enemas. A degree of constipation is to be expected in a patient in pain and lying motionless, but it is likely that a large iliacus haematoma has a more direct effect on the bowel.

Ecchymosis—We have not seen skin discoloration in the vicinity of the haematoma, but Tallroth (1939) mentioned it.

Age incidence—Iliacus haematoma appears to be unique among the haemorrhagic accidents of the bleeding diseases as it occurs mainly in adolescents and young men (Fig. 2). It is strange

that the complication should arise so rarely in the first ten years of life when haemarthroses are so frequent and when intramuscular haematomas at other sites are common.

ANATOMICAL CONSIDERATIONS

The syndrome described is the result of haemorrhage into a closed compartment, the walls of which are formed by bone and fascia and which contains the femoral nerve and the iliacus muscle. Figure 3 shows the iliac fascia in a fresh cadaver. The fascia is firmly attached at its margins to the inner lip of the iliac crest and to the brim of the true pelvis. It is connected to the posterior margin of the inguinal ligament where it is continuous with the fascia transversalis. Over the upper and lateral parts of the iliacus muscle and over all but the lowest part of the psoas muscle the fascia is thin and translucent but it is particularly strong over

FIG. 3

The posterior wall of the left iliac fossa of a fresh cadaver.

the groove between these two muscles where its transverse fibres are reinforced by bundles running longitudinally. The fascia becomes progressively thicker as it passes downwards behind the inguinal ligament and at this site forms a dense and indistensible funnel enclosing the lower parts of both muscles. The dissection in Figure 4 shows the fibres of the lowest part of the fascial sheath arching over the femoral nerve as it lies in the intermuscular groove. **Experiment**—In a fresh cadaver it was found that very large quantities of water could be injected into the sheath of the psoas muscle which distended easily in its upper part (Fig. 5). When fluid was injected into the substance of the iliacus muscle a tense globular swelling occurred, similar to the iliacus haematoma palpated clinically (Fig. 6). When further fluid was forced in it “overflowed” into the psoas sheath, which then filled up from below so that eventually there were two swellings with a deep groove between them (Fig. 7). The psoas and the iliacus muscles lie therefore in separate compartments except where they pass together into the thigh.

In iliacus haematoma haemorrhage occurs into the iliacus muscle causing distension of the thinner parts of its fascia and the rounded tumour which was felt in all our patients. As

the small iliacus compartment is not distensible, blood is forced into the dense funnel of fibrous tissue below and compresses the femoral nerve. Should bleeding continue, the psoas sheath becomes distended producing a fusiform tumour which was felt in three of our patients.

The lateral cutaneous nerve of the thigh and the genito-femoral nerve, though closely related to the muscles, lie outside the fascial envelope and therefore usually escape compression (Fig. 3).

As has been shown, iliopsoas haemorrhage is usually associated with femoral nerve palsy; yet we have not found a single record of a psoas abscess which caused nerve compression. Tallroth (1939) remarked on this. He believed that it could be explained by the relatively slow distension of the psoas sheath which occurs when a tuberculous abscess forms, and that

FIG. 4

The posterior wall of the right iliac fossa of a fresh cadaver. The fascia has been dissected off the muscles in the upper part to display the femoral nerve lying surrounded by fat in the intermuscular groove. The nerve and the iliacus and psoas muscles pass below under a dense fibrous arch.

inflamed fascia is eroded and ruptures long before the pressure within the abscess can compress the nerve. However, these considerations hardly explain the absence of nerve compression when the psoas muscle is the site of an acute non-tuberculous abscess as described by Zadek (1950). His six patients were found at operation to have tense abscesses within the psoas sheath; yet none suffered a femoral nerve palsy.

We believe that the explanation is to be found in the results of the experiment just described. The significant difference between an iliacus haematoma and a psoas abscess, whether acute or chronic, is that the former is primarily a haematoma of the iliacus muscle which may overflow into the psoas sheath.

Primary distension of the psoas sheath, from whatever cause, would be unlikely to give rise to nerve compression, because the thin, distensible fascia can contain a large volume of fluid at low pressure.

FIG. 5

FIG. 6

FIG. 7

Figure 5—Injection of the psoas sheath. Figure 6—Initial effect of injection of the sheath of the iliopsoas muscle. Figure 7—Further injection produces overflow distension of the psoas sheath.

PATHOLOGY

The morphology of iliopsoas haematoma is shown in Figure 8. The specimen consists of the left half of the sacrum and a portion of the ilium of a patient of twenty-three years with haemophilia who developed bilateral iliopsoas haematomata shortly before his death (Hall 1961). At necropsy the haematomata were found to extend from the diaphragm to the inguinal ligament. The cavity of the left haematoma communicated with the lumen of the colon. The fibro-osseous compartment which limits the haematoma is well shown. The iliac vessels, lying in front of the iliac fascia, are displaced forward by the haematoma but not compressed. It is difficult to distinguish any structures in the substance of the haematoma, but histological section confirmed that the femoral nerve traversed the cavity, completely surrounded by blood clot. In this patient the haematoma had ruptured into the bowel and had become infected. The iliopsoas muscle, remnants of which are seen compressed against the ilium, was largely destroyed by infection.

It might be supposed that haemorrhage into the fascial envelope of the muscle, in addition to compressing the nerve, would cause ischaemic necrosis of the iliopsoas muscle, yet in our cases there was no evidence that this occurred. The striking flexion of the hip which is so characteristic of the syndrome in the early stages is the result of spasm of the muscle; in two of our patients the hip extended fully under anaesthesia. Later the flexion contracture was due to the great distension of the fascial sheath and not to ischaemic contracture of the contained muscle, for although it persisted for many weeks, full extension was in every case regained when the haematoma finally resorbed and in no case was the power of flexion at the hip impaired.

DIFFERENTIAL DIAGNOSIS

In nearly all patients the presence of a severe bleeding diathesis is diagnosed before the age of three. As has been mentioned, iliopsoas haematoma occurs in adolescence and adult life and it follows that the patient will be a known sufferer and will almost always show some

evidence of haemorrhages in the past, a fact which should make misdiagnosis unlikely if the clinician is aware of the frequency of the complication. The occurrence of the syndrome in a patient undergoing anticoagulant therapy suggests that the complication should be borne in mind in acquired as well as hereditary coagulation defects.

Acute haemarthrosis or suppurative arthritis of the hip bears a resemblance to this syndrome, but the demonstration of tenderness and a mass in the iliac fossa, and the presence of a nerve lesion should distinguish them. Acute haemarthrosis of the hip is uncommon in haemophilia.

Retroperitoneal haematoma is a diagnosis commonly made in haemophilia. The term should be reserved for bleeding into the retroperitoneal space which lies anterior to the

FIG. 8

An iliacus haematoma. The left half of the sacrum and ilium of a patient of 23 years suffering from haemophilia.

iliopsoas fascia. This space is potentially very large, and a true retroperitoneal haemorrhage produces an entirely different clinical picture. A diffuse mass may be palpable, often extending into the loin, but it is not like the tense well defined tumour of an iliacus haematoma. Femoral nerve compression does not occur and there is no flexion contracture of the hip.

When it occurs on the right side, iliacus haematoma may closely mimic acute appendicitis and the diagnostic error may have dangerous consequences (Fraenkel 1957). If there is any evidence of femoral nerve compression (the earliest sign being hypoaesthesia of the skin over the patella) the possibility of appendicitis can be excluded. If nerve compression is absent, or if its onset is delayed, the diagnosis may be in doubt. Flexion deformity of the hip is more severe in iliacus haematoma than is seen in acute appendicitis. Iliacus haematoma cannot be palpated in the early stages because of intense "guarding" of the abdominal muscles, but when in doubt the iliac tumour is easily felt by examination under anaesthesia.

TREATMENT

The aim of treatment in the acute phase is to stop bleeding and relieve pain and lessen nerve damage, by correcting the clotting defect and by immobilising the affected iliacus muscle. Pain that continues in spite of correction of the clotting defect ceases when the patient is immobilised.

The patient with haemophilia is given fresh frozen plasma or a more potent concentrate of human antihæmophilic globulin if plasma proves ineffective. The patient with Christmas disease is treated with plasma, which need not be fresh but should be less than a week old. A concentrate of Christmas factor is used in a few centres for control of persistent bleeding. Infusion should be continued for at least four days to allow some organisation of the clot (Biggs and Macfarlane 1962).

Bed rest, with pillows to support the affected hip in flexion, may be adequate in slight hæmorrhage, but when bleeding continues and pain is severe immobilisation should be complete. A modified Robert Jones frame has been used with success in several cases.

Once the pain has subsided the flexion contracture diminishes, and its correction may be helped by light traction to the leg. Protective infusions may be given again for a few days when mobilisation and weight bearing are begun. When walking is allowed it is important to protect the knee with a caliper and to continue this protection until the quadriceps femoris muscle has regained full power. If this precaution is omitted recurrent hæmarthrosis of the knee is almost inevitable. We have examined many patients in whom the only residual disability from an iliacus hæmatoma was a disorganised knee from repeated hæmarthroses which damaged it beyond repair during the months of quadriceps paralysis.

PROGNOSIS

In the published cases we have studied, recovery of femoral nerve function has been unpredictable. Two patients made a complete recovery (Seddon 1930, Lyons 1953); three showed a considerable improvement (Günther 1924, 1935; Weil 1946); and two showed none (Tallroth 1939, Douglas and McAlpine 1956).

We have reviewed twenty cases of iliacus hæmatoma in seventeen patients a year or more after hæmorrhage. In fifteen recovery was complete or marred only by a small area of numbness over the patella. In five there was weakness and wasting of the quadriceps femoris muscle but power was not less than 4 on the Medical Research Council scale. The prognosis for nerve recovery is excellent, even in severe hæmorrhage, if proper treatment is given. In one of our patients, who suffered iliacus hæmorrhage while in hospital, it was possible to institute treatment immediately and he never developed signs of nerve compression. This is the only patient we have seen who escaped a nerve palsy and the only patient who got prompt treatment. This experience suggests that if treatment is given at once nerve palsy may be avoided or at least diminished.

SUMMARY

1. Hæmorrhage into the fascial compartment which contains the iliacus muscle and the femoral nerve is a common complication of hæmophilia.
2. The iliacus hæmatoma syndrome is described and illustrated from the authors' study of thirty episodes occurring in twenty-four patients.
3. The anatomy of the iliopsoas fascia is described and the mechanism of femoral nerve compression explained.
4. Differential diagnosis, prognosis and treatment are discussed and the necropsy findings in one patient are presented.
5. An instance of iliacus hæmatoma occurring as a complication of anticoagulant therapy is recorded.

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