Musculoskeletal images. Desmoplastic fibroma of the thigh

A 25-year-old woman presented with a 2- to 3-month history of increasing discomfort in the left anterior hip and pelvic area. She had also noted a decrease in her stride length, as well as a nontender mass in the thigh. There was some dysesthesia in the leg with long periods of sitting. She denied suffering any associated injury. Her medical history included a protein-S deficiency, a recent atypical Papanicolaou smear that precipitated a cone biopsy of the cervix, and a recent intentional 4-kg weight loss. There was a strong family history of malignant disease.

On physical examination her gait was normal and she could squat fully. There was a mobile, firm, slightly tender, soft-tissue mass, 9×13 cm in dimension, palpable in the anteromedial aspect of the proximal left thigh. Range of motion in her hip was restricted to 115° of flexion versus 140° on the normal right side. Internal and external rotation was possible to 40° and 45°, respectively, both abduction and adduction to 50°. She had tight hamstrings, a negative Lasèque test and a negative bowstring sign. Findings on neurosensory examination were normal. There was a 3.5×5.5-cm geographic café-au-lait spot on the medial thigh, and similar ones on the right lower abdomen, left arm, posterior right thigh and posterior left thigh. There was no other evidence of neurofibromatosis.

Plain radiographs of the pelvis with inlet and outlet views revealed a 3.0-cm radiolucent defect in the left inferior pubic ramus (Fig. 1). There was some disruption of the inferior cortex, where a 2.0- to 3.0-cm fragment had been avulsed from the inferior aspect, probably at the adductor insertion. There was increased density in the adjacent soft tissues. There was no calcification within the lesion. The appearance of the pelvis was otherwise normal. The overall appearance was that of a nonaggressive process.

Computed tomography (CT) of the pelvis, enhanced with contiguous 10-mm scans, showed evidence of a fairly extensive heterogeneous soft-tissue mass within the proximal adductor compartment of the left thigh, with resultant loss of definition of intramuscular septal planes and expansion of the corresponding adductor component. The mass also demonstrated discrete, poorly defined punctate ossification and calcification suggestive of chondrocalcification. It abutted on the inferior pubic ramus and appeared to extend from an inferiorly directed pedunculated exostosis.

Magnetic resonance imaging (MRI) of the left thigh revealed an expansile, 3.2×1.2-cm lesion in the inferior pubic ramus. It had low signal intensity, similar to adjacent muscle on T_1-weighted images. The bony cortex was disrupted at the inferior aspect and was associated with a minimal amount of periosteal new bone formation. Immediately adjacent to, and inseparable from, the lesion at its lateral and inferior aspect, was a large associated soft-tissue mass measuring $8.6 \times 7.9 \times 9.5$ cm (Fig. 2). The mass bulged slightly through the obturator foramen and was inseparable from the lateral fibres of this muscle. The pectineus muscle was draped over the superior aspect of the muscle mass, and the epicentre of the mass was in the adductor compartment. The posterior margin of the mass was inseparable from the quadriceps muscle. The mass lay just below the left femoral head and left hip joint with no evidence of extension into the joint or of a joint effusion. The mass was separated from the femoral vessels by adductor muscles and the proximal left femur. There was no definite evidence of a calcified matrix. After gadolinium was administered, there was slight heterogeneous enhancement of the lesion.

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An open biopsy revealed a bland, hypocellular spindle cell lesion with no mitotic activity or necrosis. The features were consistent with a desmoplastic fibroma of bone with extensive soft-tissue involvement or an extra-abdominal fibromatosis or desmoid tumour of the thigh with secondary bone involvement.

En bloc excision of the soft-tissue mass along with resection of the inferior pubic ramus was performed. An inguinal ventral hernia was closed, and the resultant defect in the left groin and thigh was repaired with a pedunculated mini-tram transverse abdominal flap.

Gross examination of the excised specimen showed a 10.5 × 8.5 × 7.5-cm circumscribed mass involving the bone of the inferior pubic ramus and the adjacent soft tissues of the thigh. The sectioned mass was firm and white, with a whorled and microcystic appearance (Fig. 3). Microscopic examination revealed hypocellular, bland, spindle cell proliferation, with low mitotic activity and no necrosis (Fig. 4). Collagen deposition, small cystic spaces and focal myxoid areas were seen. No malignant osteoid was present. Although the edges of the lesion were infiltrative, there was no lymphvascular or perineural invasion, and the resection margins were free of tumour. On immunohistochemical staining, the tumour cells stained with vimentin and, focally, for smooth-muscle actin. Testing for cytokeratins, S-100 protein, desmin, epithelial membrane antigen and CD34 was negative. Cytogenetic analysis revealed a normal 46,XX karyotype. The features were those of a desmoplastic fibroma of bone.

Desmoplastic fibroma is a rare primary bone tumour first described by Jaffe in 1958. It is characterized by a locally aggressive spindle cell tumour that resembles the pattern of fibromatosis in soft tissues. It occurs typically in adolescents and young adults. The long bones and mandible are most commonly affected, but the pelvis may also be involved. Pain and swelling are the most common presenting symptoms. The classical radiographic appearance is that of an expansile, lytic lesion with well-defined

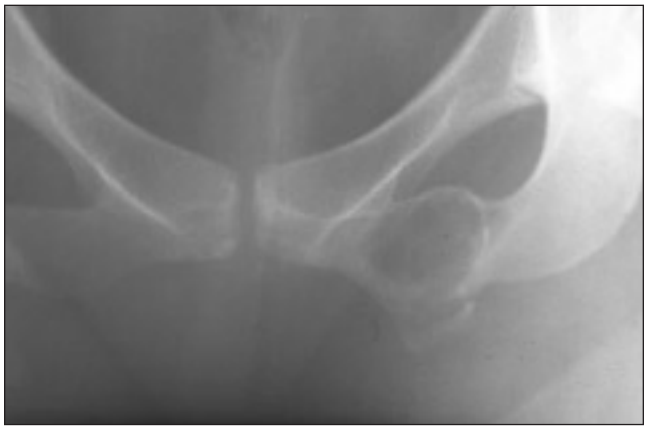

FIG. 1. Radiograph of the affected area of the left thigh shows a 3.0-cm radiolucent defect in the left inferior pubic ramus with increased density in the adjacent soft tissue. There is no evidence of calcification within the lesion.

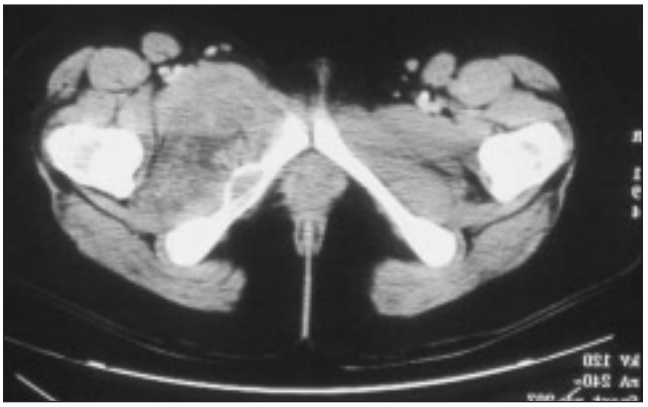

FIG. 2. T_1-weighted magnetic resonance image shows an expansile lesion in the inferior pubic ramus with an associated large soft-tissue mass.

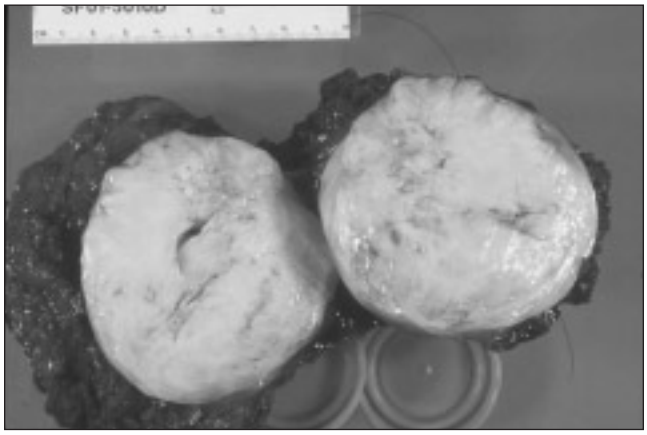

FIG. 3. The tumour is circumscribed, with a white-grey, firm and whorled appearance.

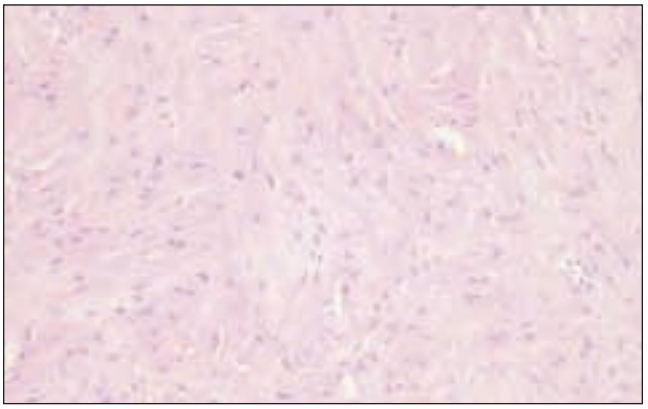

FIG. 4. Bland spindle cells in a collagenous stroma are typical of desmoplastic fibroma of bone (hematoxylin–eosin; original magnification ×20).

margins and little, if any, periosteal reaction. Destruction of overlying cortex with extension into the soft tissues is not uncommon. CT and MRI are useful to delineate the lesion and its relationship to adjacent structures. Tumours can range in diameter from 3 to 20 cm and have a grey-white, fibrous, solid appearance. Microscopically, desmoplastic fibroma is similar to soft-tissue fibromatosis, consisting of spindle-shaped fibroblasts, dense collagen matrix, rare mitotic figures and variable cellularity. The lesion exhibits an infiltrative, destructive pattern with permeation of bone marrow spaces and haversian canals, as well as surrounding soft tissues. Ultrastructural and immunohistochemical studies reveal prominent myofibroblastic differentiation.

The clinical behaviour of desmoplastic fibroma is similar to that of aggressive fibromatosis of soft tissues in that local infiltration and recurrences are common, but metastatic disease does not occur. Simple curettage and bone grafting can result in a 40% recurrence rate, so wide local excision is the recommended treatment.■

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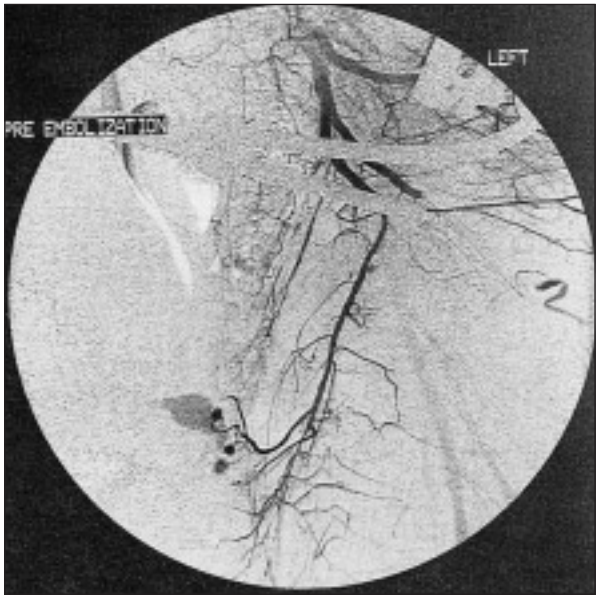
Category 6, Items 36 and 37



Pelvic fractures are common after high-speed motor vehicle crashes. Successful treatment requires rapid resuscitation from hemorrhagic shock, identification of associated injuries, and bony stabilization to reduce long-term disability.

Based on the extent of injuries, celiotomy is necessary. In the case described, the laceration of the perineum is diagnostic of an open fracture and extends into the rectum. To prevent continued fecal contamination of the pelvic spaces and hematoma, a diverting colostomy with distal colon washout is indicated. Debridement, irrigation, and cleansing of the contaminated area are warranted, and packing of the area may be needed to control bleeding. External pelvic fixation closes the pelvic space and is effective in control of bleeding in most cases. Continued pelvic bleeding is a life-threatening problem, but operative ligation of the hypogastric vessels in an attempt to decrease pelvic bleeding is not useful because of extensive collateral flow, and because most bleeding originates from the fracture sites. Operative ligation of these vessels would require opening the contained pelvic hematoma, leading to further hemorrhage.

After external pelvic fixation and in the absence of another obvious source for bleeding, continued hypotension and transfusions are indicative of pelvic vascular injury. Angiography is indicated to locate and embolize the source of the bleeding (see angiogram). This usually stabilizes the patient, assuming all other sources of bleeding have been addressed. A repeat CT scan will not localize the site of injury and celiotomy with pelvic packing is not effective in controlling arterial bleeding. Application of a pneumatic antishock garment (PASG) is not indicated because it severely limits patient access, at times results in extremity ischemia from compartment syndrome, and is not effective in controlling arterial bleeding. Diuretic therapy is not indicated for hemorrhagic shock.

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