Fibrous Dysplasia of the Spine

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Fibrous dysplasia affecting the spine is unusual. A further 11 cases are reported and the radiological features are described. The complications and difficulties with the diagnosis of this condition are discussed.

In 1938, Lichtenstein introduced the concept of polyostotic fibrous dysplasia to describe a number of cases in which the medullary cavities of the involved bones were replaced by fibrous tissue containing primitive bone trabeculae. In a survey of the disorder in 1942, Lichtenstein and Jaffe described such lesions of single or multiple bones with a predominantly unilateral involvement. They also described the association of the more severely affected cases with abnormal skin pigmentation, precocious puberty, hyperthyroidism and other extra-skeletal abnormalities.

Fibrous dysplasia may affect any bone. Polyostotic lesions are most commonly found in the femur, tibia, pelvis and humerus, monostotic lesions in the femur, tibia, ribs, maxilla and mandible (Gibson and Middlemiss, 1971). Involvement of the vertebral column is uncommon and is usually associated with polyostotic fibrous dysplasia; only six cases of vertebral involvement in the monostotic variety have been reported (Resnick and Lininger, 1984).

This article describes the radiological features of a further nine cases of vertebral involvement in polyostotic fibrous dysplasia and another two cases of the monostotic form.

Table 1 – Radiological appearances and sites of involvement of vertebral lesions in nine patients with polyostotic fibrous dysplasia.

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Site of involvement</th>
<th>Radiological features</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Cervical spine</td>
<td>Expansion of the laminae and spinous processes of C1 and C2 with a ground glass appearance.</td>
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<tr>
<td>2</td>
<td>Cervical spine</td>
<td>Expansion of the right transverse process of C7 by a lucent lesion with some apparent internal septation.</td>
</tr>
<tr>
<td>3</td>
<td>Cervical spine</td>
<td>Collapse of the body of C2 with expansion of the posterior elements with lucent areas. Possible expansion of the spinous processes of C3 and C5 (Fig. 1).</td>
</tr>
<tr>
<td>4</td>
<td>Thoracic spine</td>
<td>Collapse of the right side of the vertebral body of T9 with extrusion of the body anteriorly. Also expansion of the right transverse process of T10 by a lesion with well defined margins and some internal septation probably extending into pedicle (Fig. 2).</td>
</tr>
<tr>
<td>5</td>
<td>Lumbar spine</td>
<td>Minor expansion of the body of L2 by a fairly well-defined lucent lesion with an incomplete sclerotic rim (Fig. 3).</td>
</tr>
<tr>
<td>6</td>
<td>Lumbar spine</td>
<td>Loss of height antero-inferiorly of the vertebral body of L2. Lucent lesion within body with sclerotic margin and some internal septation. No involvement of posterior elements. The L2–3 disc space is narrow and L3 has remodelled to fill the gap (Fig. 4).</td>
</tr>
<tr>
<td>7</td>
<td>Lumbar spine</td>
<td>Well-defined lucent lesion with some expansion of L2 vertebral body and an incomplete sclerotic rim. Possible extension into pedicle.</td>
</tr>
<tr>
<td>8</td>
<td>Lumbar spine</td>
<td>Expansion of involved left pedicle of L3 (Fig. 5).</td>
</tr>
<tr>
<td>9</td>
<td>Sacral spine</td>
<td>Expansion of right lateral mass of S1 with sclerotic rim and lucent centre (Fig. 6).</td>
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Fig. 1. – Involvement of the body of C2 has resulted in collapse with a localised kyphosis.
Fig. 2 - Collapse has occurred of the right lateral aspect of the ninth thoracic vertebral body. The tenth thoracic vertebra is also involved with expansion of its right transverse process and pedicle.

MATERIAL AND METHODS

The material was drawn from the radiological library of the Institute of Orthopaedics at the Royal National Orthopaedic Hospital, Bolsover Street, London and reviewed by two radiologists (D.J.S. & J.F.C.W.).

RESULTS

Nine cases of spinal involvement in patients with polyostotic fibrous dysplasia and a further two cases of monostotic vertebral involvement are reported. In the polyostotic cases, typical radiological appearances of fibrous dysplasia were present in other bones. In both

Table 2 - Radiological appearances and sites of involvement of vertebral lesions in two patients with no other evidence of fibrous dysplasia

<table>
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<tr>
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<tbody>
<tr>
<td>10</td>
<td>Cervical spine</td>
<td>Marked expansion of the spinous process of C2. A lucent lesion with a well defined cortical margin and some internal septation. Biopsy proven fibrous dysplasia (Fig. 7). Expansion of the lamina of C5 with a thick sclerotic rim surrounding a well defined rounded lucent area containing a central dense nidus. The appearances were consistent with an osteoblastoma. Fibrous dysplasia was proved on biopsy (Fig. 8).</td>
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<tr>
<td>11</td>
<td>Cervical spine</td>
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</table>

Fig. 3 - A well-defined lesion of L2 is present with some expansion of the vertebral body; the rim of the lesion is sclerotic in places.
Fibrous dysplasia of the spine

Fig. 4 – Involvement of the second lumbar vertebral body by a lucent lesion with a sclerotic margin and some internal septation is shown. A long-standing collapse of the inferior vertebral plate has resulted in reduced height of the disc and remodelling of L3.

Monostotic cases the diagnosis was confirmed histologically. The distribution and radiological features are summarised in Tables 1 and 2.

DISCUSSION

Fibrous dysplasia may be classified into several types: (a) monostotic; (b) pauciostotic, with lesions similar to the monostotic form and not showing the general involvement of the polyostotic form, but with several bones involved in one region, e.g. involvement of the pelvis and proximal femur; (c) polyostotic; and (d) involvement of the facial bones, which is sufficiently different to merit separation into a distinct type.

Involvement of the spine in fibrous dysplasia is uncommon (Epstein, 1976; Murray and Jacobson, 1977; Resnick and Lininger, 1984). Its frequency varies in different series. Some report no spinal involvement (Stewart et al., 1962; Reed, 1963; Gibson and Middlemiss, 1971) but in others vertebral involvement occurs in 7% to 24% of patients (Harris et al., 1962; Henry, 1969; Warrick, 1973). Vertebral involvement in fibrous dysplasia shows the same features as lesions in the appendicular skeleton: (a) expansion of bone, sometimes with thinning of the cortex; (b) central, mainly lytic, lesions of the vertebral body or appendage, with well-defined sclerotic margins, and (c) generalised mixed lytic and sclerotic involvement of a vertebral body, sometimes giving the appearance of coarse trabeculation. A soft tissue mass may be present.

When involvement of the vertebral body becomes extensive, vertebral collapse may occur, usually as a consequence of a compression fracture of one or other vertebral plate (Grabias and Campbell, 1977). Eight
Fig. 6 – Frontal radiograph of the sacrum which shows expansion of the right lateral mass of the first sacral segment. The lesion has a sclerotic rim and lucent centre.

Fig. 7 – The spinous process of the second cervical vertebra is markedly expanded. The lesion has a well defined margin with a lucent centre and some internal septation.

Fig. 8 – Lateral tomography demonstrates expansion of the lamina of the fifth cervical vertebra. The lesion has a thick sclerotic rim surrounding a well-defined rounded lucent area and a central density.
instances of spinal cord compression have been described (Rosencrantz, 1965; Montoya et al., 1968). These were not always related to the vertebral collapse but were sometimes produced by a posteriorly expanding fibrous tissue mass. Cord compression may also be caused by expansion of either the vertebral body itself or of pedicles and articular processes.

In most cases of fibrous dysplasia, the regions of lesser density are not completely radiolucent only appearing so in comparison to the surrounding sclerosis of the normal cortical bone. In fact, comparison of such areas with the adjoining normal vertebral medulla indicates that density usually is increased overall. This is the 'ground glass' appearance of fibrous dysplasia in other locations, and is due to the presence of many irregular spicules of bone within the fibrous stroma.

As fibrous dysplasia of the spine is uncommon and solitary lesions are often asymptomatic, vertebral lesions in the monostotic disease are the least common manifestations, discovered as incidental findings during radiological examination for some other reason. However, when such lesions occur and are not diagnosed radiologically, the absence of a characteristic lesion elsewhere may necessitate further investigation and even biopsy before the diagnosis can be established. In this series one monostotic lesion was mistakenly identified as an osteoblastoma, and in other series vertebral involvement has been said to resemble non-ossifying fibroma, Paget's disease, aneurysmal bone cyst, hyperparathyroidism, and vertebral plana due to eosinophilic granuloma (Warrick, 1973; Gualtieri et al., 1978).

Book Review


This book contains the proceedings of a workshop held in Angers on 25–28 November 1986, and addresses a vital aspect of radiation protection which judging by the lack of published data in recent years, has received far less attention than it deserves. The 45 papers in this book address the subject which has now established for itself as a routine diagnostic tool. Although this book is invaluable for the radiation protection specialist, it is not cheap, and it suffers from the disadvantages of presentation that S-factors are being developed for the 3, 6 and 9 month pregnant woman to allow for such displacements. This book contains no information to fill the yawning gap in the data describing human placental transfer of radionuclides and their biokinetic behaviour in pregnant women and in children, despite the firm place which paediatric nuclear medicine has now established for itself as a routine diagnostic tool.

The Chernobyl accident has heightened the need to provide age-related radiation protection standards and has provided the unwelcome opportunity of obtaining human data. Two papers in this book relate somewhat unusual ways in which human data has been obtained: Rondo (UK) traced the Cs-137 activities from long range weapons fallout in his own children throughout the mid 1960s, and Schlenker and Keane (USA) assayed the skeletal Ra-226 and Ra-228 in children, despite the firm place which paediatric nuclear medicine has now established for itself as a routine diagnostic tool.

REFERENCES


