Odontoid fracture complicating Jaffe–Lichtenstein disease

Case report

Johann Peltier, M.D., Michel Lefranc, M.D., Anthony Fichten, M.D., Carole Cordonnier, M.D., Patrick Toussaint, M.D., Ph.D., Christine Desenclos, M.D., and Daniel Le Gars, M.D., Ph.D.

1Service de Neurochirurgie and 2Laboratoire de Cytologie et d’Anatomie Pathologique, Centre Hospitalier Universitaire d’Amiens Nord, Amiens, France

The authors report the case of a 31-year-old man with polyostotic fibrous dysplasia who suffered a traumatic odontoid fracture in an area of a preexisting bone lesion. Conservative treatment was successfully performed. Jaffe–Lichtenstein disease is discussed along with differential diagnoses and alternative methods of treatment.

(DOI: 10.3171/SP/2008/8/295)

Key Words • Jaffe–Lichtenstein disease • odontoid fracture • polyostotic fibrous dysplasia

Fibrous dysplasia of bone is a rare benign disease characterized by bone pain, various fractures, and bone deformities that can be extremely debilitating. It can occur in 1 bone (monostotic form) or several bones (polyostotic form, also called Jaffe–Lichtenstein disease). The spine, especially the superior cervical section, is rarely affected. To our knowledge, we report the third case of axial involvement in fibrous dysplasia and the second case of odontoid fracture.

Case Report

History and Examination. This 31-year-old man had received a previous diagnosis based on biopsy results of polyostotic fibrous dysplasia (Fig. 1). He presented with a complaint of acute neck pain following an automobile accident. He was a passenger in the back seat and had been wearing a seat belt. Since the age of 21 years he had had a history of Jaffe–Lichtenstein disease (polyostotic fibrous dysplasia). At that time he had undergone surgery for a left ptertrochanteric femoral fracture (Fig. 2). His family history was unremarkable.

In the emergency room, this patient was found to be neurologically intact. There was no retropharyngeal hematoma. A physical examination revealed only pain located at the cervicooccipital joint and nuchal rigidity. Moreover, he had been walking with a cane for the past 3 years and had developed progressive left hypoacusis. Neither café-au-lait spots nor physical stigmata of endocrinopathy were noted on admission.

The erythrocyte sedimentation rate, and serum concentrations of parathyroid hormone, 25-hydroxyvitamin D, serum phosphate, and calcemia were all within normal limits. Only the alkaline phosphatases were abnormally high (817 IU/L; normal range 41–117 IU/L). This patient was treated by intravenous pamidronate with calcium and vitamin D every 6 months.

Plain radiographs of the upper cervical spine revealed increase density within the axis but were difficult to interpret. A cervical CT scan clearly demonstrated a pathological fracture through the odontoid base (Type II). The fracture line was slightly oriented toward the bottom and the front with an anterior displacement of < 2 mm. We noted that the posterior arch of the axis was dysplastic with a higher degree of bone condensation (Fig. 3). Also a posterior fusion by placement of a C1–2 Apofix construct (Medtronic Sofamor Danek) was not possible. Cerebral and thoracic CT scans showed skull base and costovertebral involvement (Fig. 4).

Treatment. We elected to treat our patient conservatively because the fracture line was near anatomical structures. In addition, the involvement of the C-2 posterior arch did not allow performing a safe posterior fusion between either C-2 and C-1 or between the C-1 and C-3. Finally, Jaffe–Lichtenstein lesions, as with Paget disease, are very hemorrhagic during surgery and can cause significant blood leakage. This patient was maintained with a sternococcipitomandibular immobilizer brace for 3 months, and a good result was attained (Fig. 5). The patient also was not treated with administration of intravenous pamidronate because he had no phosphate wasting on urinary measurements.
Postoperative Course. Follow-up frontal and sagittal CT scanning performed 9 months after the injury demonstrated union of the dens to the body of the axis (Fig. 6).

Discussion

Fibrous dysplasia of bone or Jaffe–Lichtenstein disease is a rare but potentially severe skeletal disorder characterized by malfunction of bone-forming mesenchyme. There are 2 clinical forms of this disease: the polyostotic form affecting multiple bones, particularly long bones; or the monostotic form involving a single bone. Fibrous dysplasia accounts for 2.5% of all bone lesions and 7% of benign bone tumors. McCune–Albright syndrome is a type of fibrous dysplasia in which polyostotic lesions are accompanied by abnormal melanotic cutaneous macules called café-au-lait spots, precocious sexual development, and other endocrinological abnormalities. This syndrome appears to be more frequently found in females.

Involvement of the vertebral column is anecdotic and is mostly seen in the polyostotic form (at a rate of 4–14% of cases) than in the monostotic variety. Involvement of the upper cervical spine occurs in 1.4% of cases. The majority of cases involving the spine are discovered incidentally on radiography because the lesions are usually asymptomatic. Fibrous dysplasia affects males and females equally, manifests predominantly during childhood, adolescence, or pregnancy, and often becomes quiescent after skeletal maturation during adulthood to remain clinically silent or rarely regress. Sarcomatous transformation (osteosarcoma, fibrosarcoma, or chondrosarcoma) is very uncommon and occurs in 0.4–6.7% of cases.

Histologically, fibrous dysplasia is a benign lesion characterized by replacement of the medullary component of bone with collagenized fibrous tissue and irregular osteoid formation, whereas the cortex is always intact but thin and spongy. This fibrous dysplasia of bone within the interwoven fibrous tissue has an appearance often likened to alphabet soup. Cytological atypia and mitosis are not observed.

The clinical presentation in 77% of cases is pain progression and rarely myelopathy or radicular sensory and/or motor loss. The median age at time of diagnosis is 18 years, and the median age at onset of treatment is 28 years. In the monostotic form the dysplasia can remain clinically silent into adulthood and is often revealed incidentally on radiographs. At the time of diagnosis, one quarter of patients have a prior history of trauma. Only 0.7% of patients have neurological deficits. Ossicular and cochlear impingement may develop during adulthood.

In laboratory investigations, phosphaturia which is commonly associated with polyostotic fibrous dysplasia is likely to contribute to the propensity of lesional bone to fracture.

Radiologically, CT scans reveal a very thin, osteolytic, bubble-like cortex sometimes accompanied by widening of
abnormally mottled bone or liquid contents.\textsuperscript{14,23,30} In addition, there are granular vertical striations of the vertebral body or radiolucency.\textsuperscript{12,18,28,36,38} Involvement of both the vertebral bodies and posterior elements has been reported in 75\% of patients, and isolated anterior disease has been reported in 20\%.\textsuperscript{26} Bone lesions have been reported in 90\% of cases of unilateral disease and even in cases of bilateral involvement, the lesions tend to occur on 1 side.\textsuperscript{5,9,12,13} On magnetic resonance imaging, T1-weighted images show fibrous dysplasia as hypointense, and T2-weighted images reveal fibrous dysplasia as either hypointense or hyperintense and there is neither significant intraosseous hemorrhage nor associated soft mass tissue.\textsuperscript{4,17,28,37} Finally, 3 radiographic patterns have been identified: pagetoid, sclerotic, and cyst-like.\textsuperscript{10}

The differential diagnosis at the time of imaging often includes osteoblastoma, vertebral hemangioma, eosinophilic granuloma, aneurysmal bone cyst, giant cell tumor, chor- doma, and solitary metastasis in monostotic dysplasia form.\textsuperscript{14,15,26} For instance, solitary eosinophilic granuloma of C-2 involves the ventral area (vertebral body) and is usually lytic with preservation of vertebral height, little paravertebral reaction without disc involvement.\textsuperscript{24} A CT scan of a multiple myeloma of the axis demonstrates disruption of the anterior and posterior cortical surfaces at the origin of the odontoid process with important demineralization.\textsuperscript{25,27}

The treatment of fibrous dysplasia consists of administration of intravenous pamidronate, 60 mg for 3 consecutive days, every 6 months with calcium and vitamin D supplements to avoid secondary hyperparathyroidism. Indeed, at commencement of treatment, 5\% of patients present with secondary hyperparathyroidism induced by changes in dysplastic bone in adults with elevated serum parathyroid hormone secondary to vitamin D deficiency.
nate belongs to bisphosphonates drugs, which inhibit bone resorption and osteoclastic activity. These cycles are repeated for 2 years and after those 2 years, pamidronate is reserved only for patients with imaging-documented improvement and pain disappearance. This protocol displays a 41% reduction in the pain intensity, whereas 95% of patients with a dens fracture in multiple myeloma experience relief of their pain with radiotherapy alone. A favorable radiological response was reported to have been seen in 54%, characterized by filling of the lytic lesion and/or cortical thickening. Undesirable effects are transient fever and venous irritation.

In cases of spinal involvement, no consensus of management has been contraindicated. Of fractures in polyostotic fibrous dysplasia, 79% are treated conservatively (traction and/or casting) and 21% are treated surgically. The healing potential for all fracture sites has been reported to be 94%. The indications for surgery depend on the topography, lesion size, amount of displacement (significant displacement when > 5 mm), symptoms, and deformity (significant degree of angulation when > 10°). Apuzzo et al. showed that Type I fractures (through the upper part of the odontoid) and Type III fractures (that extend into the body itself), fractures displaced < 4 mm, and fractures in patients < 40 years of age are thought to be likely to heal with conservative treatment. A significant displacement may disrupt the arterial arcade around the dens formed by both vertebral arteries and compromise...
Odontoid fracture in fibrous dysplasia

vascularization of the bone elements of the dens, thus predisposing to rate of nonunion.35

Conclusions

Involvement of the axis in Jaffe–Lichtenstein disease is very uncommon. Our case poses only the problem of therapeutic planning because the diagnosis of the polyostotic form was known since young adulthood. The successful treatment of the patient requires recognition of preexisting bone lesions and thorough anatomical study of the superior or cervical spine. Periodic follow-up examinations are necessary to check safe healing.

References

15. Ledoux-Lebard G, Souljin C: [Vertebral localization of fibrous dysplasia of bone, or Jaffe-Lichtenstein disease; two case reports.] J Radiol Electrol Arch Electr Medicale 34:349–353, 1953 (Fr)
18. Lichtenstein L, Jaffe HL: Fibrous dysplasia of bone: a condition affecting one, several or many bones, the graver cases of which may present abnormal pigmentation of skin, premature sexual development, hyperthyroidism or still other extraskeletal abnormalities. Arch Pathol 33:777–816, 1942


Address correspondence to: Johann Peltier, M.D., Service de Neurochirurgie, Centre Hospitalier Universitaire d’Amiens, Place Victor Pauchet, 80054 Amiens cedex 1, France. email: jojo.peltier@caramail.com.

J. Neurosurg.: Spine / Volume 8 / March 2008