Fibrous dysplasia of the atlas

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Fibrous dysplasia (FD) is a localized, benign progressive replacement of bones by a fibrous proliferation intermixed with poorly formed trabeculae of woven bone. Polyostotic FD commonly involves the femur, skull, tibia, humerus, ribs, fibula, radius and ulna, mandible, vertebrae, and other bones. The vertebral involvement accounts for only 2.5% of cases. On literature review, we found only 27 reported cases involving the cervical spine, and most of these cases presented with pain syndrome or radiculopathy in the sub-axial vertebrae. The aim of this article is to present a case of FD involving the atlas that presented with myelopathy.

We report a 16-year-old male patient, who presented to our institution complaining of occipital pain, which radiated to the right upper extremity for the last 3 years. This pain was persistent and relieved temporarily by analgesics. Six months prior to presentation he started to have a progressive right upper extremity weakness, which was worse distally with an inability to write anymore. This weakness was associated with unsteady gait and no fecal or urinary control problems. He sought medical advice from different institutions and was referred to our service later. On examination he was found to be conscious, alert, and showed no cranial nerves abnormalities. The motor power was 3/5 distally, and 4/5 proximally in his right upper extremity; otherwise, the power was normal in the rest of the limbs. He had spasticity in both legs and exaggerated reflexes all over his extremities, with positive plantar reflexes, and sustained clonus. The sensory examination was normal. A CT of the cervical spine (**Figure 1**) showed diffuse thickening and expansion of the atlas, forming an exophytic lesion on the right side that measures 4.5 x 4 x 3.5 cm at the anteroposterior, transverse, and craniocaudal dimensions respectively. This lesion compromised the neural canal with significant spinal cord compression. Another lesion at the fifth vertebral body was seen that carried the same radiological characteristics as the atlas lesion, but without a mass effect (not shown). A CT angiogram showed occlusion of the right vertebral artery. An MRI of the C-spine showed a bony lesion at the atlas level with significant spinal cord compression. A C-spine dynamic view showed no evidence of instability at the craniocervical junction. Liver function test, serum thyroxine level, estradiol, follicle-stimulating hormone (FSH), leuteinizing hormone (LH) levels, growth hormone, adrenocorticotropic hormone, and cortisol levels were all within normal values. He underwent decompression of the posterior spinal cord through a C1 posterior arch piece meal laminectomy with preservation of the facet capsules. Intra-operatively, the posterior arch of C1 was found to be hypertrophied with pseudospinous process formation. The bone was extremely vascular but controllable with bone wax. Post-operatively, he was kept on mechanical ventilation overnight and was extubated the next day, where he was transferred to the surgical ward in a stable condition. He started to improve dramatically regarding the weakness and myelopathy. Post operative CT scan showed a satisfactory posterior decompression of the neural canal. He was discharged home on day 5 post operatively, and was seen in the outpatient department 2 months later. He continued to improve with right arm power of 4+/5 and steady gait. A year and a half later he was asymptomatic with full functional recovery. Microscopic examination showed the lesion to be composed of irregular fragments of woven bones, some of which were rimmed by well-defined osteoblasts. Occasional osteoclasts were seen. The background stroma was hypocellular and composed of bland spindle cells. Neither atypia nor mitotic figures were seen. The described zonation of this lesion was not appreciated in our specimen because of its fragmented nature.

Fibrous dysplasia is a genetic disorder, caused by a mutation in the GNAS1 gene that encodes the alpha subunit of the stimulatory G protein. There are 4 possible presentations of FD. 1) Involvement of a single bone (monostotic form). 2) Involvement of

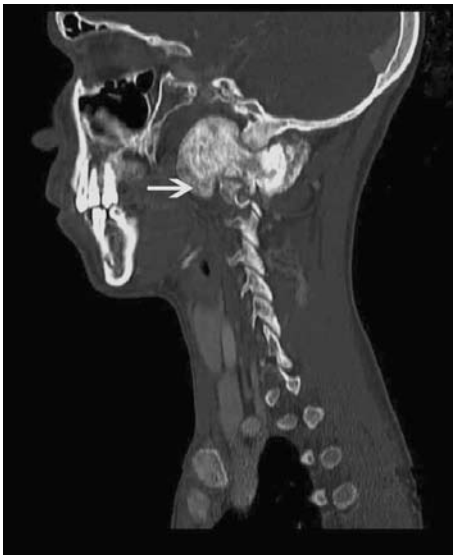

Figure 1 - Sagittal reconstruction of cervical spine presenting the extensive bony hyperplasia of the arches of the atlas (arrow).

several bones in different regions (polyostotic form). 3) Involvement of several bones in one region (pauciostotic form), and 4) FD as part of another syndrome, such as, Mazabraud syndrome in which FD is associated with intramuscular myxomas, or McCune Albright syndrome, which is a polyostotic disease associated with various endocrinopathies and skin pigmentation. Our patient is a pauciostotic FD since there is involvement in C1 and C5. Management of cervical spine involvement is uncertain, as the preoperative diagnosis is usually difficult using the non-invasive modalities. In our case, he was showing slow progressive compression of the spinal cord. A percutaneous biopsy seemed unnecessary, as he required surgical decompression. The 2 main concerns were spinal cord compromise and craniocervical junction stability. Through the dynamic views there was no evidence of subluxation, which would change the surgical goals and strategy.

On review of the previous reported cases of cervical FDs, instability was a main issue and was treated conservatively through external arthrodesis or surgically through instrumented fusion.[1] There is one mortality reported from failure of the fusion. The bone graft can be involved in the FD process. We elected to decompress the posterior arch only and follow up the patient clinically and radiologically through repeated CT scans. The possible outcomes of this approach are spontaneous fusion of the craniocervical junction as part of the natural history, stabilization of the disease process as reported before, or progression of the disease to recurrent myelopathy with or without instability. In either case, another surgery would be necessary to decompress and possible stabilize the patient.

The possible pathological complications are bone cyst formation that can predispose to fractures,[2,3] and rare malignant transformation (fibrosarcoma). Thus, a long-term follow up is crucial in this case. Intravenous pamidronate appears to induce a marked decrease in pain severity, with improvement of bone strength and radiological appearance, and a decreased risk of fracture through bone resorption inhibition. This can be considered as a medical option for pain control in our case, as the pain can be the only presenting symptom,[4] however, pain was not a major issue to the patient. Van Giffen et al[5] reported a C1 fibrous histiocytoma involving the posterior arch in a 6-year old boy. In their report, the main presentation was pain syndrome in the same location as our case, but less extensive involvement, however, surgical intervention was necessary in both cases. A complete resection was feasible in Van Giffen et al's case due to limited involvement of the atlas.

In conclusion, cervical spine FD is a rare condition. It has a clinical spectrum of presentation ranging from asymptomatic to myelopathy. Surgical decompression and close follow up can be the main stay of treatment.

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References

1. Marshman LA, David KM, O'Donovan DG, Chawda SJ. Fibrous dysplasia of the cervical spine presenting as a pathological fracture. ***Br J Neurosurg*** 2004; 18: 527-533.
2. Lomasney LM, Basu A, Demos TC, Laskin W. Fibrous dysplasia complicated by aneurysmal bone cyst formation affecting multiple cervical vertebrae. ***Skeletal Radiol*** 2003; 32: 533-536.
3. Dang D, Baig MN, Christoforidis G, Chiocca EA, Gabriel J. C2/C3 pathologic fractures from polyostotic fibrous dysplasia of the cervical spine treated with percutaneous vertebroplasty. ***Eur Spine J*** 2007; 16 Suppl 3: 250-254.
4. Sambasivan M, Sanalkumar P, Mahesh S, Nair KR, Basheer A. Monostotic fibrous dysplasia of the C4 cervical spine. ***Neurol India*** 2008; 56: 497-498.
5. van Giffen NH, van Rhijn LW, van Ooij A, Cornips E, Robben SG, Vermeulen A, et al. Benign fibrous histiocytoma of the posterior arch of C1 in a 6-year-old boy: a case report. ***Spine (Phila Pa 1976)*** 2003; 28: E359-E363.