

Monostotic Fibrous Dysplasia of Thoracic Spine

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SUMMARY

Monostotic fibrous dysplasia of the spine is rare. We describe a case report of 13+8 year old girl who experienced low thoracic back pain while doing back flips in gymnastics class and subsequently a tingling sensation, progressive paraplegia, clonus, and the development of hyperactive reflexes in her lower extremities. She underwent successful anterior and posterior spine stabilization surgery along with the removal of the tumor and was able to return back to full sports activities, including gymnastics. The research work was conducted during my post graduate fellowship training at Minnesota Spine Centre USA under the supervision of Dr. Francis Denis (an author of Denis three column theory of spine).

Keyword: Monostotic fibrous dysplasia of the thoracic spine-progressive paraplegis, tumor excision, spine stabilization.

INTRODUCTION

Fibrous dysplasia generally is considered to be a developmental anomaly characterized by the failure of differentiation of bone forming mesenchyme⁵. Fibrous dysplasia may be confined to a single bone (monostotic) or to the bone of a single extremity (monomelic) or may be distributed through several or many bones throughout the body (polyostotic). The fundamental process is proliferation of cellular fibrous tissue which destroys normal cancellous bone and marrow elements, and encroaches upon (and expands the cortex in which characteristic trabeculae of metaplastic bone form). This condition usually is identified in infancy or early childhood and is thought to be congenital¹⁴.

Fibrous dysplasia accounts for 2.5% of all bone masses and 7% of all benign bone tumors²³. It can involve any part of the skeleton. The lower extremity and pelvis are more commonly involved followed by the ribs and upper extremities^{1,3,5,6,36}. When long bone is involved it includes the metaphyseal area but rarely the entire length of bone. Epiphyseal plate involvement is very rare. Spine involvement also is relatively uncommon especially for monostotic types^{1,8}. Skeletal involvement can be monostotic (70-80%) or polyostotic (20-30%). Monostotic types occur more frequently than polyostotic^{5,6,8,13,15,33,34} although the polyostotic form is much more commonly reported in literature, probably because of its severity.

The characteristic clinical presentation of fibrous dysplasia is pain or pathological fracture^{8,13,25}. The radiological appearance is usually typical showing expansive trabeculated lesions with preserved

cortical margins^{13,20,25}. Differential diagnosis between fibrous dysplasia and other isolated lytic lesions is critical. Excisional biopsy and histopathological examination is necessary to make the final diagnosis⁸.

On the basis of different reports, it has been estimated that polyostotic fibrous dysplasia involves the cervical spine (7%) and lumbar area (14%) although the true incidence probably is much lower^{4,7,10,23}. However, no exact percentage can be quoted about the monostotic form, as it is rarely reported in literature.

The first case of monostotic fibrous dysplasia involving the thoracic spine was reported by Bruce, et al. in 1987²³. There are few other reports regarding spine involvement by fibrous dysplasia in different areas. We present a different case of monostotic fibrous dysplasia of thoracic spine with secondary elements of aneurysmal bone cyst.

CASE REPORT

This is a case of a 13+8 year old female, 5'6" in height and 98 pounds in weight, who reported immediate onset of low thoracic back pain while doing back flips in a gymnastic class, and started having tingling sensations in her lower leg with gradual loss of ability to balance and walk straight over the following two weeks. She saw her local physician and was worked up with plain film computerized tomography (CT) scan and a magnetic resonance imaging. A T10 lesion was found, and she was referred to the Minnesota Spine Center for further treatment on April 22, 1992. Approximately six weeks after the initial episode. On physical examination her spine was not tender to palpation and sensation was diminished to pin prick at

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approximately T11 to T12, levels with loss of proprioception. Motor examination revealed 3/5 iliopsoas, 4/5 quadriceps, 4/5 hamstrings, 3/5 gastroc, 4/5 tibialis anterior, and 3/5 EHL, with deep tendon reflex 3+P at the knees and ankles along with clonus and upgoing Babinski's. Good rectal tone with voluntary control was noted. Her AP and lateral radiographs showed evidence of obliteration of the right T11 pedicle with a lytic defect. CT scan showed a lytic defect involving all the posterior elements of T10 into the pedicle, particularly the right side. Both the anterior and posterior aspect of the vertebral body appeared to be intact. There was significant canal stenosis which was confirmed by results of MR imaging.

Surgical Technique

The patient underwent an anterior spine fusion T10-T12, posterior spine decompression, tumor excision and spine fusion from T10 to T12 with Cotrel-Dubousset instrumentation 2 days after for initial exam at the Minnesota Spine Center. A 10th rib thoracotomy approach was utilized by a thoracic surgeon. This provided access to the T10-T11 and T10-T12 disc spaces. The disc spaces were removed, endplates decorticated, and morselized rib grafts were packed into the interspaces. No lesion on the anterior aspect of the bone was observed. The patient was then turned prone on a Hall-Ralton frame, prepped and draped in a sterile fashion. Subperiosteal dissection was carried out from T10 to T12. The tumor was identified and the outer layers of the tumor mass were removed. The outer layers of the tumor mass were found to be quite cystic with both cystic and solid portions. The T11- T12 interspace was entered first as it was less tight than the T10-T12 interspace. Tumor tissue was removed. A cervical Kerrison was used to remove a portion of the superior lamina of T12 to gain access to the epidural space. The tumor material was delivered out of the epidural interspace, grasped anteriorly and released. A Leksell rongeur was used to debulk the mass of the tumor and then careful blunt dissection with a nerve hook was used to free up tumor tissue from the epidural space. The superior lamina of T12 was thinned out down to the epidural space with a carbide and then a diamond bur. Dissection was accomplished with a nerve hook to free up the tumor tissue, a cervical curet to release, and a Kerrison to remove loose debris. The entire tumor was removed up to the pedicle of T10. CD rods, hooks, and screws were placed posteriorly with no correction performed.

Operative Findings

There was no lesion on the anterior aspect of the bone. Posteriorly there was a large expansile type lesion containing both cysts and firm whitish tumor tissue. The dura was very compressed by the

expansile tumor. The frozen section was found to have a lot of spindle cells, and eosinophilic granuloma was suspected. Specimens were sent for Gram's stain, culture and sensitivity. Aerobic and anaerobic, fungal culture, TB culture-all were reported negative.

Pathology Report

On gross examination, the pathologist reported 1.8 x 1.2 x 0.8 cm. pale tan firm centrally cystic mass; three fragments of pale tan firm tissue, the largest being 1.2 x 0.8 x 0.3 cm. and approximately 3 grams of multiple fragment of pale round to pale tan firm calcified material consistent with fragment of bone, the largest being 1.7 x 0.4 x .2cm.

Microscopic examination showed that the lesion tissue was composed of a dense fibrous spindle cell stroma containing irregular, round to oval shaped osteoid arranged in a nonfunctional manner. Features which are typically seen in fibrous dysplasia. The other area showed features of an aneurysmal bone cyst including cystic spaces surrounded by loose fibroblastic septae containing a few multinucleated giant cells and osteoid production. So, on the basis of the operative and pathology findings, the diagnosis of fibrous dysplasia with secondary aneurysmal bone cyst was made.

Postoperative Course

In the recovery room, the patient was able to move her lower extremities. Postoperatively the patient was continued on intravenous steroids which had been started prior to her transfer to MSC. Postop chest x-ray indicated minimal atelectasis at the left base which resolved. Her chest tube was removed on 2nd post-op day and antibiotics also were stopped. She had a VO scan six days postoperatively due to complaints of chest pain which found no abnormality and no apparent pulmonary infarct. The patient demonstrated 3/5 EHL and 4/5 strength for all other muscle groups of her lower extremities. She was then immobilized in a molded TLSO brace and was able to ambulate with some difficulty. She continued to report numbness and tingling in her leg initially, but less than experienced preoperatively. She was discharged to home six days after the surgery with a treatment plan to include physical therapy consisting of gentle parascapular and shoulder stretching, range of motion exercises, upper extremity strength program, and gradual walking training.

One month after the operation her physical examination demonstrated normal strength to 5/5 in all lower muscle groups, no clonus, and brisk but symmetrical knee and ankle reflexes. On subsequent follow up, approximately 9 months later, she showed full strength in lower extremities with normal reflex and radiographs showed solid fusion with instrumentation in good position. Six months later, at

most recent follow-up, she was allowed to return to full physical activity, and she is doing well.

DISCUSSION

Before 1937 no proper differentiation was made between monostotic and polyostotic fibrous dysplasia. In 1937 Albright, Butler, Hampton, and Smith¹ described the condition known as Albright syndrome in children, characterized by wide spread skeletal changes, cutaneous pigmentation of the Cafe-au-lait type, endocrine disorders as shown by sexual precocity with early secondary sex characteristics and (rarely) thyroid malfunctions. They also noted mental deficiency in few patients with extensive bony involvement¹⁷. In 1942 Lichtenstein and Jaffe first recognized and differentiated the monostotic and polyostotic pattern of fibrous dysplasia^{15,30}.

Early efforts were directed at describing the pathological and radiological appearance of the disease, and it was then demonstrated that monostotic and polyostotic fibrous dysplasia are indistinguishable histologically^{8,30}. In 1951 Strausburg, Garber, and Hallock called attention to progressive radiological changes in fibrous dysplasia³³ in the beginning there are areas of decreased density within the cortex that have a "ground glass" appearance, usually at metaphyseal bone, later increasing in size, expanding and thinning the cortex, and finally producing local osteoporosis with subsequent bowing. Robb-Smith (Falconer, Cope et al. 1942) described this process as "a primary collagenous osteogenesis of the marrow with attritive osteolysis of the lamellar trabecular". Strassburg, Garber et al³³ reported that there is progressive maturation of fibrous dysplasia tissue, that it begins as proliferation of connective tissue in the medullary space which encroaches upon and destroys cancellous trabeculae, subsequently expanding and eroding the cortex. In the monostotic form it continues to proliferate until patients grow Harris, et al⁶ reported that in his series, 56% of the patients had extension of lesion over time Hennery documented radiographic progression of the disease with pregnancy^{7,30}. The radiological picture of the lesion varies from purely lytic, to a classical ground glass appearance, to sclerotic or densely calcified. Moreover it is difficult to diagnose monostotic fibrous dysplasia by radiological pattern only³⁶ and biopsy is usually necessary for confirmation^{3,8,15,30}. Lytic lesion, haemoangiomas, giant cell tumor, aneurysmal bone cyst with blastic lesion, Pagets disease, and osteoblastoma, all should be included in a differential diagnosis.

In the majority of patients, the lesion of fibrous

dysplasia becomes clinically evident in the first two decades of life and the patient may present with a variety of manifestations. Typically, painless bony swelling and pathological fracture is seen. Fascial involvement with resulting bony enlargement may produce asymmetry, nasal obstruction or cranial nerve compression, visual difficulties, facial paralysis and ansomnia². Moreover, the patient may present with weakness of extremities or even with paraplegia, as reported in this case.

Whether the lesion of fibrous dysplasia resolves or becomes active with the onset of puberty is still controversial. Reed (1963)²¹ noted no change in the biopsy specimen that were obtained over a period as long as ten years. Similarly Harris⁷ et al. reported that the examination of serial biopsy specimens demonstrated only subtle histological differences over time. They observed decreased cellularity and slight decreased quantity of bone relative to fibrous tissue, but no Conversion of fiber-bone trabecular to lamellar bone.

In far less than one percent, fibrous lesions have been known to develop into fibrosarcoma, osteosarcoma, chondrosarcoma or even giant cell tumors²⁹. The literature is sparse on monostotic fibrous dysplasia involving the spine and only eleven cases have been reported.

Schulumberg (1946)²⁵ described 67 cases of monostotic fibrous dysplasia with only one patient having spinal involvement (cervical). Rosendahl-Jenson (1956)²⁴ and Resencrantz (1965) from Europe each described an isolated case with cervical (C4) and thoracic spine involvement. Ledoux-Lebard and Souldquin (1953)²³ reported two cases involving the L1 and L4 vertebral bodies. Harris⁷ in 1962 reported a case involving the transverse process of the 4th lumbar vertebra. Alfred Khan and Paul Rosenberg (1980) reported monostotic fibrous dysplasia involving the pedicle and the body of the 3rd lumbar vertebra. Resnik and Lininger²² (1984) described vertebral involvement, and Bruce, Rosenberg, Christopher et al²³ in 1987 reported another case involving the vertebral body, right pedicle, superior articular facet and lamina, with superior extension of the mass to the C7 right inferior facet⁸. Finally, in 1992, Shigeru, Susan, and Andrew reported two cases of monostotic fibrous dysplasia involving lateral masses of C1 and the body of the L3 vertebra respectively.²⁶ Clearly there appears to be no predilection for any particular level of spine, with both vertebral body and posterior element involvement possible.

Monostotic fibrous dysplasia does not exhibit the laboratory abnormalities, extra skeletal manifestation, or endocrinopathy^{4,7,9,23,24,31,36} seen in the polyostotic form. Its diagnosis is often difficult and usually is

based on confirmation by histopathological report. The characteristic "ground glass" appearance is not always diagnostic as other lesions can also present in the same way. Magnetic Resonance Imaging is the modality of choice, as it best demonstrates the characteristics of lesion¹⁸. Normally the cortical bone of the spine is a signal voided region. The pathological bone of fibrous dysplasia has higher signal intensity than the normal medullary bone of the vertebra due to replacement of mature bone with irregular woven bone and fibrous tissue and compression of normal medullary bone. The heterogeneity of the high intensity signal is secondary the interwoven elements which occur in this process^{18,23}. Other tumors of the vertebral column more typically present homogeneous appearance and frequently demonstrate a lower intensity signal than the surrounding medullary bone¹⁷. Computed tomography also provides additional usefull localizing information.

Both tests are helpful in ruling out an aggressive process with cortical destruction or soft tissue extension. Radio nuclei bone scan studies reveal an increased uptake the fibrous dysplasia lesion and are as useful as a screening test to rule out other sites of skeletal involvement².

Surgical treatment of fibrous dysplasia is advised if there is neural compression which may be due to collapse of the vertebral body, or to expansion of either the vertebral body itself or the pedicle and transverse process. The posteriorly expanding fibrous tissue mass also can compress the neural element. In this case, early surgical intervention was recommended because of the presence of neurological symptoms, the location of the lesion with respect to the structural stability of the spine, and to some extent, the uncertainty of the diagnosis.

CONCLUSION

Monostotic fibrous dysplasia involving the thoracic spine is rare. Only two cases have been reported in the literature. We describe a rare case of fibrous dysplasia with secondary aneurysmal bone cyst. We emphasize that when making a diagnosis of fibrous dysplasia, the possibility of aneurysmal bone cyst, giant cell tumor, haemangioma paget disease, osteoblastoma, hyperparathyroidism (Brown tumor) and eosinophilic granuloma should be kept in mind. In the presence of neural compression, early surgical excision of the tumor, and spine stabilization, is recommended in order to avoid permanent damage to the cord by the lesion.

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