Fibromyxoma of the Axis

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Abstract

Fibromyxoma of bone is a rare benign tumor of fibrous tissue origin. The typical location is the jaws. Sporadic extragnathic cases have been reported, but fibromyxoma of the spine has not been reported. The histological appearance of fibromyxoma is benign and includes abundant extracellular fibrous and myxoid stroma with varying amounts of calcification and ossification. Myxoid changes are usually extensive. Extragnathic fibromyxoma of bone should be distinguished from benign cartilage-forming bone tumors, such as chondromyxoid and myxoid chondrosarcoma and myxoma of bone. It has also been suggested that fibromyxoma is a variant of myxoid fibrous dysplasia, whereas other authors reported extragnathic fibromyxoma resulting from myxoid degeneration of bone tumors, such as chondrosarcoma or fibrosarcoma. The overtreatment of patients with fibromyxoma of bone due to an aggressive imaging appearance should be avoided; the prognosis is excellent compared with the jaw variant and depends on the location and extent of the tumor.

This article describes a case of a 21-year-old woman with fibromyxoma of bone originating from the spinous process of the axis. Clinical examination showed a tender mass in the midline of the posterior aspect the neck and slight limitation of neck range of motion; neurologic examination was normal. Diagnosis was obtained with a preoperative biopsy. Marginal excision of the lesion with posterior laminectomy of the axis was performed. The facets were preserved, and no fusion was performed. At last follow-up 2 years after diagnosis and treatment, the patient was asymptomatic with no evidence of local recurrence.

Figure: Lateral radiograph of the cervical spine showing no evidence of local recurrence 2 years after diagnosis and marginal tumor excision with posterior laminectomy of the axis.

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Fibromyxoma of bone is a rare benign tumor of fibrous tissue origin. The typical location is the jaws.\(^1\)\(^-\)\(^4\) Compared with gnathic tumors, extra gnathic fibromyxoma usually shows benign proliferation and biologic behavior.\(^3\)\(^-\)\(^10\)

Since its original description,\(^4\) sporadic extragnathic cases have been reported.\(^1\)\(^-\)\(^4\)\(^,\)\(^7\)\(^-\)\(^17\) Predilection sites are the metaphyses of the long bones. Locations in flat and nontubular bones are rare.\(^2\)\(^,\)\(^12\)\(^,\)\(^17\) Fibromyxoma of the spine has not been previously reported. This article describes a case of a patient with fibromyxoma of bone arising at the posterior arch of the axis.

**CASE REPORT**

A 21-year-old woman presented with a 5-month history of neck pain and tenderness. She reported no neck injury, and her medical history revealed episodes of cervical pain treated conservatively over a 4-year period. Clinical examination showed a tender mass in the midline of the posterior aspect of her neck and slight limitation of neck range of motion. Neurologic examination was normal.

Cervical spine radiographs showed a tumor originating from the spinous process of the axis, extending to the fifth cervical vertebra (Figure 1). Trocar biopsy histology showed a lesion composed of a shell of compact bone rimming wide medullary spaces with fibrous and adipose tissue. Scattered spindle cells with slightly hyperchromatic nuclei set in an edematous to myxoid matrix existed in the fibrous component. Several ectatic vessels filled the medullary spaces (Figure 2). No atypical mitosis or infiltration was observed. The histological diagnosis of fibromyxoma of bone was made, and the patient underwent excision of the lesion with posterior laminectomy of the axis.

Intraoperatively, the tumor originated from the spinous process of the axis. It had a smooth surface, a bilobate shape, and a maximum diameter of approximately 7 cm. The facets were preserved, and no fusion was performed. Histology of the excised specimen confirmed the preoperative diagnosis; the surgical margins were marginal but microscopically negative. One month postoperatively, the patient reported mild pain and normal range of motion of the cervical spine. Cervical spine radiographs 2 years after treatment showed no evidence of local recurrence (Figure 3).

**DISCUSSION**

The rare occurrence of fibromyxoma of bone impedes classification and distinction of this tumor from other fibrous tumors containing focal myxoid tissue.\(^8\)\(^,\)\(^14\)\(^,\)\(^15\)\(^,\)\(^18\)\(^-\)\(^20\) Fibromyxoma should be distinguished from benign cartilage-forming bone tumors, such as chondromyxoid and myxoid chondrosarcoma based on the absence of a histological lobular pattern, chondroid matrix and variety of cells, and the more aggressive behavior of myxoid chondrosarcoma.\(^1\)\(^,\)\(^3\)\(^,\)\(^4\)\(^,\)\(^8\)\(^,\)\(^14\)\(^,\)\(^20\) Fibromyxoma is also similar to myxoma of bone.\(^3\)\(^,\)\(^9\)\(^,\)\(^10\) However, myxomas are characterized by the proliferation of primitive mesenchymal cells that produce amorphous intercellular substance.\(^3\)\(^,\)\(^19\) In addition, patients with myxomas are older than 60 years, whereas patients with fibromyxomas have an even age distribution. In older patients, more myxoid matrix is a possible sign of tumor
regression. It has also been suggested that fibromyxoma is a variant of myxoid fibrous dysplasia characterized by exuberant mucin production and growth potential. However, the immature fibrous tissue and the typical osteoid of fibrous dysplasia are not found in fibromyxoma.

Aneurysmal bone cysts should also be screened microscopically for preexisting fibromyxoma, especially in patients younger than 40 years. Other authors regarded extragnathic fibromyxoma as resulting from myxoid degeneration of bone tumors, such as chondrosarcoma or fibrosarcoma.

Pain is the typical clinical presentation of patients with fibromyxoma of bone. However, various benign and aggressive imaging patterns have been reported. The most common radiographic patterns are a well-defined radiolucent lesion with cortical destruction without a periosteal reaction, an expansile central metaphyseal lesion with a sclerotic rim, and a malignant lesion with ill-defined margins, cortical destruction, and soft tissue extension. An extrasosseous mass has been reported in approximately one-third of cases. Because of the variable radiographic appearance, differential diagnosis of fibromyxoma is difficult, and the definite diagnosis is histological.

The histological appearance of fibromyxoma is benign and includes abundant extracellular fibrous and myxoid stroma with varying amounts of calcification and ossification. Myxoid changes are usually extensive. The cells are spindle shaped or stellate depending on the myxoid stroma; mitosis and pleomorphism is not observed. Blood vessels exist in the tissue but are not numerous.  

**CONCLUSION**

Complete curettage or excision of fibromyxoma is the treatment of choice. A local recurrence rate of 13% has been reported after incomplete excision. Cryosurgical techniques as adjuvants to curettage eliminate the risk of local recurrence and may be beneficial if long bone segments are involved. Overtreatment because of an aggressive imaging appearance should be avoided. The prognosis is excellent compared with the jaw variant and depends on the location and extent of the tumor. Metastases from fibromyxoma of bone have not been reported.

**REFERENCES**