Desmoplastic Fibroma of the Skull; A Case Report, Review of the Literature, and Therapeutic Implications

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= Abstract =

두개골에 발생한 결합조직성 섬유종 증례보고 및 문헌 검토

Desmoplastic fibroma (DF) is a rare neoplasm of the bone, and is histologically benign but locally aggressive disease. A total of nine cases of DF involving skull have been reported in the literature, and among these eight are females. In this report, the clinical findings and histopathology of a case with DF of the skull in a male patient is presented with a review of the literature with an emphasis on treatment modalities. A 21-year-old man presented with headache. CT scan revealed a solitary and lytic skull lesion without brain invasion. DF was confirmed by histological evaluation. On immunohistochemical staining of the tumor was negative for estrogen or progesterone receptors. After total resection of tumor with wide surgical margin, there was no recurrence during the 35 months of follow-up period. Although longer follow up period maybe needed, treatment of this type of tumor with complete resection of tumor tissue along with a wide margin may provide long disease-free state compare to the high recurrence rates in DF of other sites.

KEY WORDS Desmoplastic fibroma· Parietal bone· Operative treatment.

Introduction

Desmoplastic fibroma (DF) is a rare bone tumor. It is histologically benign but locally aggressive intraosseous neoplasm that was initially described as a distinct clinicopathological entity by Jaffe in 1958. It accounts for only nine of the 8542 osseous tumors reviewed at the Mayo Clinic. Sites of predilection are mandible, long bones, and iliac bone. Only 9 cases of DF have been reported involving skull, all except one in female patients.

In this report, we present the radiographic and histopathological findings of a case with DF in the right parietal bone of a male patient.

Case Report

A 21-year-old Korean male patient was initially evaluated for persistent right-sided headache that had lasted several weeks prior to admission. He had no history of head injury or other medical illnesses. Physical examination revealed a painful protruding mass on the right parietal region. Routine skull x-ray showed a round 4-cm lytic lesion in the right parietal bone without bone expansion or sclerosis (Fig. 1). Computerized tomography (CT) demonstrated a lytic skull lesion with destruction of both skull tables (Fig. 2). The lesion had a central soft-tissue component without brain invasion.
The skull lesion was completely removed along with wide margin and a cranioplasty was performed. Gross examination of the specimen revealed a firm fibrous gray tumor between the internal and external tables of the skull. Microscopic examination revealed that the lesion was composed of relatively uniform spindle-shaped fibroblasts with neoplastic proliferation and with interspersed inflammatory cells (Fig. 3, Right). There was infiltrative growth of tumor tissue into the bone with entrapped sclerotic bony trabeculae (Fig. 3, Left). These findings were consistent with DF. Immunohistochemical stains for estrogen and progesterone receptors were negative. There was no evidence of recurrence during the 35 months of follow-up period.

Discussion

Desmoplastic fibroma, a very rare benign tumor occurring predominantly in the metaphysis of long bones. This case adds to the nine cases of DFs in the skull that have been previously reported and is the second case which occurred in a male patient (Table 1).

Bhm reviewed 171 cases of DF in 1996 and found that the numbers of male and female patients were equivalent with a mean age at presentation of 23 years. However, DFs that have been previously reported in the skull were predominantly in females with female to male ratio of 8:1 and the patients’ ages ranged from 7 years to 86 with a mean age of 36 (Table 1). The parietal bone (4 cases) was the most frequently involved while three lesions were in the temporal bone and two in the frontal bone.

Triantafyllou et al. suggested a possible hormonal dependence of DF and reported a patient who showed a rapid proliferation of a recurrent tumor during pregnancy. The desmoid tumors of soft tissue considered as nonosseous counterpart of DF were found to express estrogen and progesterone receptors in up to two-thirds of the investigated patients. In addition, one-half of the desmoid tumors of soft tissue responded to endocrine therapy. For this reason, we performed immunohistochemical studies to verify the possible presence of estrogen and progesterone receptors. Both were negative in our case.
Various skull lesions may radiographically mimic the appearance of DF\(^1\). There are no specific radiological features of DF in that a similar radiological finding can also be seen in other lesions such as eosinophilic granuloma, aneurysmal bone cyst, medullary fibrosarcoma, hemangioma, ameloblastoma, and metastatic tumors from the thyroid or kidney\(^4\). Thus, the final diagnosis requires a careful histological examination.

Grossly, the tumor varies in color from gray to white and has a firm and rubbery consistency. Microscopically, DF is composed of mature fibrous tissue consisting of small, sparse fibroblasts with little or no mitotic activity in an abundant stroma of collagenous matrix\(^2,6,10\). This feature is important in histologically differentiating DF from low grade fibrosarcoma. The typical fibrosarcoma is more cellular with a herringbone pattern that shows more pleomorphism and higher mitotic activity. However, in some cases of low grade fibrosarcoma, mitoses may not be evident and areas with predominant collagen tissue may be found, making the distinction from DF extremely difficult\(^19\).

In such cases, the final diagnosis can be established only after follow-up evaluation\(^13\). Because of the high recurrence rate of DF (about 20-30% of all cases) and local invasiveness, complete resection with a wide margin is mandatory\(^14\). According to Pensak, hormonal regulation was considered effective in young patients and to date, early response to Tamoxifen appears to be beneficial although long-term evaluation is needed\(^18\). Previously reported cases of DF in the skull were treated by complete resection without adjuvant therapy. None of these cases showed recurrence unlike DFs of the long bones. The authors believe that this may be due to the feasibility of wide resection of skull lesions as compared to DFs of other sites.

Since our case was negative for immunohistochemical staining for estrogen and progesterone, no adjuvant therapy was warranted.

**Conclusion**

This case represents the tenth case of DF involving the
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skull. It is suggested that DF of the skull should be best treated by complete resection with a wide margin to prevent a recurrence.

References

두개골에 발생한 결합조직형성 섬유종; 증례보고 및 문헌 검토

김주한·박경윤·정용구·서중근·김성남·서연림

= 국문조목 =


중추 신경계에 발생한 경우의 위치는 다음과 같다.