Desmoplastic fibroma of the skull

Case report

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Desmoplastic fibroma is a rare fibrous bone tumor, histologically benign but locally aggressive, that was initially described as a distinct entity in 1958 by Jaffe.7 This neoplasm constituted only nine of the 8542 primary bone tumors reviewed by Dahlin and Unni.1 We have found 150 previous cases of this tumor,6 only three of which affected the skull.2,5,9 We report the radiographic and histopathological diagnosis of a case of desmoplastic fibroma involving the right parietal bone.

Case Report

This 28-year-old woman presented at our hospital complaining of continuous right-sided headaches for several weeks. Physical examination revealed a painful protrusion above the right parietal bone. Skull x-ray films showed a round 2-cm lytic lesion in the right parietal bone without sclerosis (Fig. 1). A computerized tomography scan defined the skull lesion as a lytic area involving both tables with bone fragments and a central soft-tissue component; there was no invasion of the brain (Fig. 2).

The patient underwent surgical removal of the tumor. Gross pathological examination revealed a fibrous gray-white elastic tumor between the internal and external tables of the skull. Microscopic examination showed abundant collagen fibers randomly arranged in bundles and sparsely distributed fibroblastic spindle cells, with slender nuclei (Fig. 3). A mature bone fragment was identified inside the lesion. The final diagnosis was desmoplastic fibroma. The patient has been followed with periodic examination for 3 years without evidence of local recurrence.

Discussion

Desmoplastic fibroma is a very rare benign bone tumor. Most patients with this tumor are in the first three decades of life with a peak incidence in the second decade. Pain and/or swelling and restriction of movement are frequent symptoms.3 The iliac bone, mandible, and long bones are the sites most frequently involved. To our knowledge, only three cases of this lesion involving the calvaria have been described previously.2,5,9

The radiographic findings of desmoplastic fibroma reveal lucent, localized expansile lesions with a soap-bubble appearance and mild or absent reactive bone sclerosis.4,7 Desmoplastic fibroma of the skull has been described as a solitary, purely lytic lesion without bone expansion or trabeculation. Radiographic findings are nonspecific, and several skull lesions may resemble the radiographic appearance of desmoplastic fibroma in young patients. Thus, final diagnosis requires histological examination.

Microscopically, desmoplastic fibroma is characterized by an evenly spaced population of fibroblastic-like cells embedded within a stroma containing variable amounts of collagen fibers arranged in bundles. It is important to differentiate desmoplastic fibroma histologically from low-grade fibrosarcoma; the lack of a high mitotic index, hypercellularity, pleomorphism, and other atypical cellular features in the former helps to distinguish desmoplastic fibroma from low-grade fibrosarcoma. For this reason, periodic follow-up examinations become mandatory.5

Another important feature of desmoplastic fibroma is the absence of osteoid tissue formation. In several reports,4,8 fragments of mature bone were entrapped in the periphery or scattered within the lesion, as in our case, without evidence of malignant osteoid tissue or new bone. This may result from irregular eroding and replacing of normal mature bone by the infiltrative growth of this tumor, without osseous formation.

For this skull lesion, en bloc resection with a wide margin is the treatment of choice and can be curative.7 If
Resection is incomplete, there is a high risk of local recurrence due to the tumor's local aggressiveness. In such cases, there should be periodic follow-up examinations of the patient for several years.3

References

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