Cranial chondromyxoid fibroma

Case report

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A case of chondromyxoid fibroma of the frontal bone in a 15-year-old girl is presented. This is a benign tumor, but should be excised widely enough to include a rim of normal bone, otherwise it may recur or even undergo malignant transformation.

KEY WORDS • chondromyxoid fibroma • skull tumor

CHONDROMYXOID fibroma was first established as a histopathological entity by Jaffe and Lichtenstein in 1948. The majority of these tumors arise from the long bones, but their overall incidence is low (less than 1% of all bone tumors). Those arising from the cranium are extremely rare. We could find only five reported cases of cranial chondromyxoid fibromas in the literature. To our knowledge, this is the first report to describe its appearance on computerized tomography (CT) scans.

Case Report

This 15-year-old girl visited the neurosurgery service of Wakayama Red Cross Hospital on June 6, 1980, with complaints of headache and vertigo which had lasted about 1 month, despite treatment by a local medical doctor.

Examination. She was a well developed, well nourished, healthy-looking girl. Neurological examination revealed no abnormality. Skull x-ray films showed a rounded mass in the right frontal region, which was marginally sclerotic and measured 2.5 cm in its greatest dimension (Fig. 1). The scalp lying over the mass was normal and not painful when palpated.

On admission on August 1, 1980, the patient was physically and neurologically normal. Radiologically, the cranial tumor mass had not increased in size since the previous examination 2 months earlier. Routine investigations of blood, urine, and cerebrospinal fluid were normal. Right carotid angiography demonstrated no abnormality, such as increased vasculature, around the tumor.

On CT scanning, the tumor was shown as a high-density mass on three consecutive sections, each 10 mm apart. It appeared to be protruding into the cranial cavity from the inner plate of the skull bone (Fig. 2). The CT density was high near the base and relatively low at the top of the tumor. On CT with contrast injection, the top of the tumor was enhanced. A preoperative diagnosis was made of osteogenic tumor arising from the inner plate of the frontal bone.

Operation. Cranietomy was carried out on August 13, 1980. The periosseous and the outer plate of the cranial bone over the tumor area were grossly normal. The tumor was partly adherent to the dura mater, and after separation the hemispherical tumor was excised with a rim of frontal bone which constituted the base of the tumor. The dura mater under the tumor had been thinned out, and through the dural incision the brain parenchyma was found to be molded in the shape of the tumor. Postoperatively, the patient has been doing well, and 10 months after surgery was free from symptoms.

Pathological Examination. The tumor measured 1.5 cm in diameter, and the surface was smooth and shiny. It was of rubbery consistency and uniformly pearly white (Fig. 3). On the x-ray film of the specimen, the relatively radiolucent tumor was demonstrated to be attached to the irregularly hypertrophic frontal bone.
Microscopically, the characteristic lobular pattern was noted at a low magnification. In the middle of the lobules, stellate cells with uniformly small, round-to-oval nuclei were sparsely scattered in a myxoid background, but at the periphery they are more concentrated (Fig. 4). The histology was similar in all areas sectioned, and there was no evidence of differentiation to chondroma, myxoma, or fibroma.

Discussion

Until Jaffe and Lichtenstein\(^5\) first described chondromyxoid fibroma as a separate pathological entity in 1948, it had been confused with other benign or malignant tumors of bone or cartilage. Thereafter followed other reports which, by 1970, had accumulated to 207 cases.\(^3\) The majority of chondromyxoid fibromas are seen in young people, especially in the second decade of life. Eighty percent of patients are under 30 years of age. The incidence is the same in males and females.\(^7\) These tumors are common in the metaphysial area of bones of a lower extremity, especially the tibia and femur. They are, however, extremely rare in the skull bones:\(^1,6,7,11\) we could find only five such cases in the literature.\(^1,6,7,11\) The clinical manifestations are localized headache or swelling over the tumor area, but in most cases they are of mild degree. In our case, the tumor was relatively small.
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and did not show an increase in size in the 2 months prior to surgery. There was no evidence of increased intracranial pressure, but the symptoms of headache and vertigo disappeared after the excision of the tumor.

The base of the tumor was made up of ragged bone tissue. It is unknown whether this was reactive overgrowth of skull bone or ossification of the tumor itself. The former seems more likely, because this feature was seen only at the base of the tumor, and the main mass of this tumor showed no differentiation into fibrous tissue, cartilage, or bone. The top of the tumor was not clearly seen on plain CT, but after infusion of Conray, the tumor mass was clearly demonstrated. The mean CT number for the entire tumor was 323.08 Hounsfield units ± 254.86 standard deviation (SD) on plain CT and 369.99 ± 274.30 Hounsfield units after contrast enhancement, whereas those for the central portion of the tumor excluding its margin were 194.33 ± 135.17 and 200.25 ± 108.07 Hounsfield units, respectively. The large standard deviations are probably due to heterogeneity of the tumor. Therefore, the CT number does not seem to be useful in evaluating enhancement effect in chondromyxoid fibroma.

Histologically, this tumor has a broad spectrum of components. Myxomatous areas are usually dominant, but foci of chondromatous of fibromatous areas are not uncommon. Cystic degeneration can also occur. A lobular pattern with an increased cellularity in the periphery of the lobules is an important distinguishing characteristic of this tumor. It is essential to appreciate the benign nature of the lesion and avoid unnecessary radical treatment. However, the tumor should be excised with a sufficiently wide rim of normal bone. The tumor recurred in four out of 32 cases, and even malignant transformation can follow. Effects of radiation therapy have not been evaluated.

References


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