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Chondro myxoid fibroma of skull base- A pathological dilemma

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Abstract

Background: Chondro-myxoid fibroma is relatively uncommon & is estimated to represent less than 1% of all primary bone neoplasms. Often seen in young adults during the second & third decades of life & usually arising from the metaphysis of major long bones of lower extremities. Head & Neck involvement remains a rare site & is thought to mainly arise from the synostosis at the base of skull or from sutures. This tumor may also arise from the embryonic cartilage residue. Jaffe & Lichtenstein in 1948 originally distinguished this tumor from other aggressive cartilaginous tumors. WHO defined it as a benign tumor characterized by lobules of spindle shaped or stellate cells with abundant myxoid or chondroid intercellular material separated by zones of more cellular tissue rich in spindle shaped or round cells with a varying number of multinucleated giant cells of different sizes. Establishing histological diagnosis of chondromyxoid fibroma involving skull base becomes all the more difficult because of its similarities with chondrosarcoma thus creating a trap for pathologists. Distinction between these two tumors is important due to their different management strategies. Here we present a case of chondromyxoid fibroma of right infra-temporal fossa treated by surgery alone, emphasizing more on the pathological aspects of diagnosis & differential diagnosis. A good knowledge of this clinical entity should avoid incorrect diagnosis.

Keywords

Chondro-myxoid, chondromyxoid

Introduction

Chondromyxoid fibroma (CMF) is a really uncommon tumor that addresses under 1% of all essential bone neoplasms. First depicted by Jaffe and Lichtenstein in 1948, CMFs should be recognized from other forceful cartilaginous tumors that have fundamentally various medicines and guesses. CMF is a considerate tumor described by lobules of shaft molded or stellate cells with bountiful myxoid or chondroid intercellular material with a shifting number of multinucleated monster cells of various sizes. Most much of the time, it is found in youthful grown-ups of the second and third many years of life in the lower furthest point long bones, especially emerging from the metaphysis. CMF can likewise emerge in various other anatomic destinations. Its event in the facial and cranial bones is incredibly uncommon.

CMF of the cranial bones is an incredibly provoking conclusion to make. Zilmer and Dorfman report an underlying misdiagnosis pace of 22% in their arrangement of 36 CMF cases. Contingent upon its area, CMF can be hard to recognize from an aneurysmal bone growth, sinewy dysplasia, monster cell tumor, osteoblastoma, osteosarcoma, Ewing sarcoma, mucocele, Langerhans histiocytosis, or even a schwannoma. All the more frequently, CMF is confused with three other myxoid tumors: chordoma, chondroid chordoma, and chondrosarcoma that have a more prominent recurrence of event in the craniofacial skeleton.

A 38-year-old right-gave lady gave a 1.5-year history of nasal impediment and serous rhinorrhea. Around 1 month preceding her visit, she likewise saw diplopia. A processed tomographic (CT) filter and attractive reverberation imaging (MRI) of the paranasal sinuses were acquired, which showed an expansile sore of the focal skull base, in the sphenoclival district, with augmentation into the left infratemporal fossa. An endoscopic biopsy of the mass was acquired and an underlying determination of a dangerous fibrohistiocytoma was made. On additional survey, the pathology was deciphered as showing a "Fibro-mixoid tumor, locally forceful, and with obscure metastatic potential." This patient's case was examined at our Joint Planning Conference and the suggested treatment was a medical procedure. The patient went through a transmaxillary way to deal with the foremost cranial fossa with resection of the extradurally found tumor. Postoperative course was routine and the patient was released on postoperative day 4. On follow-up, the patient detailed left facial deadness in the V2 conveyance and postoperative MRI showed lingering tumor at the left sidelong bit of the pterygopalatine gap. The last pathology report was CMF. The insignificant leftover infection was followed with sequential MR imaging. Ridiculous year, the patient's tumor showed moderate however reformist development. Careful resection of the reformist remaining sphenoclival CMF was performed. She went through a left orbitocranial way to deal with center cranial fossa with resection of the tumor. Postoperative course was dreary and the patient was released on postoperative day 2. She is sans infection at 4.5 years.

In our writing survey, we have discovered 67 distributed instances of cranial CMF since 1990. All things considered, 10 years more seasoned than those with different locales of root. Ages went from infant to 73 years. Moreover, we discovered a somewhat more noteworthy preference for CMF in females, with 35 females and 24 guys. This is clashing in the writing for certain investigations detailing a slight male inclination and others a 2:1 female-to-male event. We tracked down a slight male prevalence in the fleeting bone/occipital site subgroup. Of the cases influencing the skull, the sinonasal structures were influenced most generally, with the second most normal tumor area being the transient bone and occiput. Most tumors once in a while influenced a solitary bone, and hence seemed to develop without regard to the hard life structures by including numerous encompassing bones. Most patients were indicative at the hour of show. The guileful beginning of side effects can bring about a conceivably postponed conclusion for skull base lesions.12 Symptoms included easy or delicate growing, cerebral pain, nasal deterrent, exophthalmos, diplopia, deafness, and otalgia. The most well-known introducing manifestation for calvarial CMF was expanding. For the skull base injuries, the most well-known introducing side effects had all the earmarks of being identified with their separate areas. For

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sinonasal CMF, the most widely recognized show was nasal check, while clival/sellar sores introduced most generally with cerebral pain. Sphenoid/Parasellar injuries gave diplopia and orbital/zygomatic CMF introduced most ordinarily with exophthalmos. Last, fleeting bone/occipital CMF was most ordinarily connected with deafness at show. Given this current tumor's uncommon affinity for the calvarium and skull base, we likewise evaluated our own foundation's involvement in CMF. Since 1957 to introduce, MDACC has seen a day and a half with CMF. Of these patients, there were two cases influencing the spine: one at C2 and the other at S1/ilium. There were only one calvarial case and one skull base case,13 both announced here.

Radiologic discoveries are not indicative, but rather they can offer knowledge into the finding before intercession. Traditionally CMF is depicted as a "radiolucent, lobulated, outlined injury with a sclerotic edge and cortical development or disintegration" and calcification is uncommon. Due to the low event pace of this tumor, MRI discoveries have not been obviously settled. Be that as it may, like other bone tumors, CMF has low sign on T1-weighted and high sign on T2-weighted pictures attributable to its cartilaginous nature. The most moving viewpoint to radiologic determination of CMF is the high inconstancy of included destinations. Thusly doubt for CMF ought to be kept up while assessing singular bone injuries. Histopathologic investigation of CMF uncovers a myxoid sore containing a paucicellular focus, dull stroma cells, receptive boney spicules, and hyaline ligament with up to 75% of sores in the skull and facial bones likewise containing network calcifications. In spite of the fact that not generally present, the trademark highlights of CMF incorporate lobular appearance, chondromyxoid stroma, and sinewy tissue with multinucleated goliath cells. Nielsen et al performed ultrastructural assessment on six tumor tests discovering populaces of cells with highlights of three distinctive cell types: chondrocytes, myofibroblasts, and a combination of both chondrocytes and myofibroblasts. It ought to be noticed that if the injury shows huge atypia or mitotic action, the determination of CMF ought to be rethought. CMF is most usually discovered to be positive for vimentin, smooth muscle actin, desmin, S-100 (fluidly), and CD34. For the most part CMF is negative for pancytokeratin, carcinoembryonic antigen (CEA), and GFAP, and furthermore has a low expansion rate envisioned by Ki-67 staining.19 Veras et al portray SOX9 staining for a situation of sinonasal CMF. SOX9 has been recently portrayed as a chondrogenesis "ace controller" and assumes a part in beginning stage chondrocyte separation.