Chordoma of the posterior fossa in children

Clinical presentation is related to local invasion. Lesion of the body of the clivus can extend ventrally or dorsally and cause cranial nerve palsies, brain stem compression or hydrocephalus. Pathologically, they form soft, grey masses that are histologically benign but locally invasive and destructive. Both computed tomography (CT) and magnetic resonance (MR) imaging clearly depict the lesion. CT better demonstrates bone destruction and intralesional calcifications.

MR show all the classical signs of skull base chordoma. CT provided complementary information about bone destruction which are also usual in this type of lesion. Major differential diagnoses of the chordoma in the clivus are the other central skull base masses. Biopsy and histology make the diagnosis. Usually treatment consists in surgery and radiotherapy but more recently proton beam therapy is used and seems to yield better results [1)].

The medical records of six pediatric chordoma patients diagnosed at Childrens Hospital Los Angeles between 1995 and 2005 were reviewed. Of the six patients reviewed, all underwent an initial surgical resection. Following resection, three received a combination of chemotherapy and radiation therapy, two received chemotherapy alone and one patient refused both forms of therapy; this patient expired of progressive tumor. One patient developed acute monoblastic leukemia (M5a subtype) and died of intracranial hemorrhage during induction chemotherapy, 39 months after initial diagnosis. MRI of brain and spine showed disease progression shortly before his death. Two patients who received chemotherapy only after surgery, one patient who received chemotherapy at relapse following irradiation and one patient who received irradiation followed by chemotherapy are alive with stable radiographic abnormalities at a median follow-up of 9 years from diagnosis (range: 6-13 years). Chemotherapeutic agents included ifosfamide and etoposide in all four surviving patients. Chemotherapy with ifosfamide and etoposide may have a role in the treatment of pediatric clival chordomas when used alone or in combination with irradiation [2)].

A giant clival chordoma with disseminated disease but without involvement of the clivus. was reported as the second case, presenting without base of skull involvement, in paediatric literature and the fourth reported case of a chordoma in a patient with tuberous sclerosis [3)].

Only 6% of chordomas are localized to the cervical level. In young patients, chordomas are rare and unpredictable. Despite this, the treatment of choice remains the total resection, as much as possible, followed by proton beam radiation [4)].

1)
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