CLIVAL CHORDOMA: A CLINICAL CASE REPORT

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Summary

Introduction: The clival chordoma are rare and slow-growing embryonic tumors, originate from remains of the primitive notochord, in the center of the base of the skull with different patterns of extension in all directions, which limits the extent of tumor removal. An estimated 75% of global survival is 5 years after being treated surgically. The overall incidence is 0.08-0.5 cases per year and the location at the base of the skull is one case per 2,000,000 people. Six variants have been described: classic chordoma, chondroid, undifferentiated, sarcomatoid, intradural and extrarenal. The standard treatment is surgery; adjuvant radiotherapy is used in cases of incomplete resection; Conventional chemotherapy has not proven to be effective. Clinical studies have confirmed the efficacy of targeted therapies such as tyrosine kinase inhibitors (anti-PDGFR, anti-VEGFR or mTOR inhibitors).

Clinical case: 3-year-old girl who presented with cervical hyperextension and progressive local functional limitation that did not attenuate after symptomatic treatment with clinical progression. Subsequently, weight loss and swallowing disorder are associated; MRI showed a retro-pharyngeal tumor with extension to the cervical spine and a foramen magnum with atlo-axoid erosion. She is referred to our Center, and evaluated by the ENT, Neurosurgery and Oncology Services. An incisional biopsy was performed, which reports a classic chordoma notochordal tumor; patient receives treatment with Imatinib (2 cycles) with progression of the disease given by increase in size of the lesion, medullary bulb compression and retroclival extension, radiant treatment is decided which is rejected by the family and leaves treatment, patient dies 4 months later.

Discussion: the Imatinib indication based on clinical evidence (phase II studies), was in view of the fact that the tumor could not be surgically resected due to extension and compromise of the medullary bulb; the objective was to reduce tumor volume to subsequently apply radiotherapy, considering the age group, extensive field of radiation and sequelae after treatment.

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HOSPITAL DE NIÑOS
J.M. DE LOS RIOS

INTRODUCTION

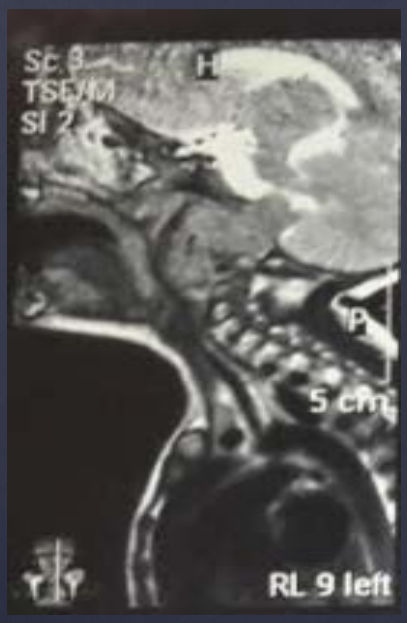
Chordomas are infrequent, slowly growing, locally destructive embryonic tumors, remnants of the first primitive originating in the center of the base of the skull with different patterns of extension in all directions, which limits the extent of tumor excision. Overall incidence is 0.08-0.5 cases per year and location at the base of the skull is one case per 2,000,000 people. 6 variants have been described: classic chordoma, chondroid, undifferentiated, sarcomatoid, intradural and extrarenal. Macroscopically, it consists of friable masses of lobular neoplastic tissue, myxoid to gelatinous appearance, bluish-white or grayish color. Microscopically a lobular pattern is observed, separated by the fibrous septa, the cells adopt a pattern of thin anastomized cords with epithelioid characteristics with broad, eosinophilic, multivacuolated cytoplasm or may contain a single vacuole that rejects the nucleus, offering an image of a signet ring cell. The symptomatology depends on the location and tumor extension, the most frequent are headache, diplopia, visual changes and paresis of low cranial nerves. The standard treatment is surgery; adjuvant radiotherapy is used in cases of incomplete resection; Conventional chemotherapy has not been effective. Phase II studies have confirmed the clinical efficacy of targeted therapies such as tyrosine kinase inhibitors (anti-PDGFR, anti-VEGFR or mTOR inhibitors), treatment with Imatinib has shown tumor stabilization (84%) or tumor size reduction in (16%) in patients with advanced chordomas in progression. The combination of bevacizumab and erlotinib shows promising results in terms of controlling tumor growth in a longer time. The survival of the untreated patients is 28 months, with a global survival at 5 years of 75% if it is treated surgically.

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CLINICAL CASE

The following is a case of a 3-year-old female patient from Edo. Apure, Venezuela, who started the current illness in December 2012 when after a bicycle crash, he presented with cervical hyperextension and progressive local functional limitation that did not attenuate after the administration of NSAIDs and rehabilitation, consulting the local physician on several occasions, with symptomatic progression. In February 2015, weight loss and swallowing disorder were associated; in MRI, a retropharyngeal tumor of approx. 2.7 x 3.4 cm with extension to cervical spine and great hole with atlo-axoid erosion, which is why it is referred to our Center, being evaluated by ENT Services, Neurosurgery and Oncology. An incisional and immunohistochemical biopsy were performed, which report a classic chordoma notochordal tumor with S-100 cytoplasmic and nuclear protein positivity.





MRI scan (october, 2015) pre-treatment

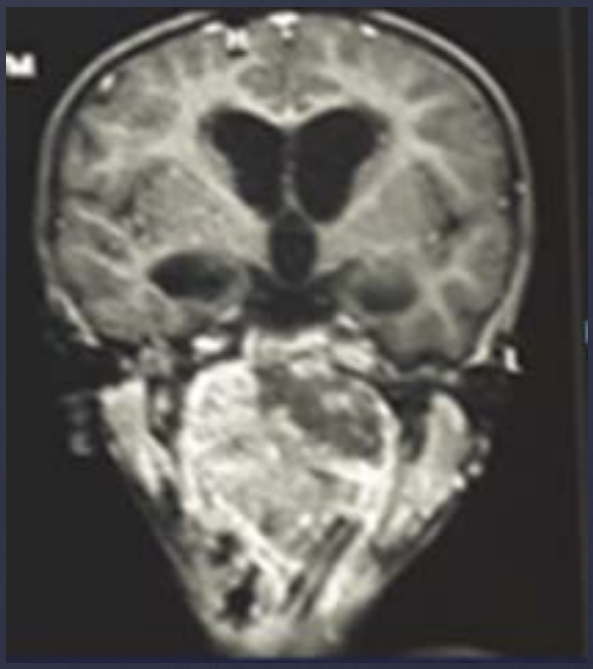
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CLINICAL CASE

Patient receives treatment with Imatinib (200 mg VO daily for 28 days) fulfilling 2 cycles with progression of the disease due to increased size of the lesion, medulary bulb compression, retroclival and nasopharyngeal extension, it is decided to indicate radiant treatment which is rejected by the patient´s family and she leaves treatment, patient died 4 months later.





MRI scan (febreuary, 2016)
Post-treatment

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DISCUSSION AND CONCLUSIONS

Chordomas are uncommon tumors (1-4% of all bone tumors), with frequent localization in the sacral (60%) and spheno-occipital (25%). With higher incidence in adults over 40 years; in children it is less than 5%, its location being more frequent in the clivus, which coincides with the case presented. Survival in untreated patients is approximately 28 months and after radiation therapy or surgical resolution is 6 to 3 years respectively. The surgical approach is the treatment of choice, which may be transsphenoidal, transfacial, transsphenoidal extended, subfrontal; remains a challenge due its rapid growth and commitment of vital structures, the most frequent complications are CSF fistulas and neuropathies. It is frequently used adjuvant radiotherapy or the indication of therapies directed as inhibitors of PDGR, EFGR (Imatinib, Cetuximab, Bevacizumab) and mTOR (everolimus / temsirolimus), because its low sensitivity to conventional chemotherapy.
In the presented case, Imatinib was chosen because it was not possible to perform tumor excision. Imatinib has more clinical evidence in terms of efficacy (phase II studies shows disease stabilization in 84% and reduction in tumor volume by 16%) and the safety profile in relation to the pediatric population. Radiotherapy was not used as a first-line treatment in view of the extensive radiation field, sequelae and age group. The treatment of recurrences is currently under study.

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