

**Cutaneous Chordoma in a Primary Localization: A Rare Diagnosis**Elatiqi Oumkeltoum<sup>1\*</sup>, Dhaidah Wafa<sup>2</sup>, Boukind Samira<sup>3</sup>, Dlimi Meriem<sup>4</sup>, MD Elamrani<sup>5</sup>, Ettalbi Saloua<sup>6</sup>, Benchamkha Yassine<sup>7</sup><sup>1</sup>Plastic surgeon, Mohammed V hospital in Meknes, morocco<sup>2</sup>Plastic surgeon, Private Practinor in Marrakech, Morocco<sup>3</sup>Plastic surgeon, Tetouan Hospital, Morocco<sup>4</sup>Plastic surgeon, Mohammed V hospital in Meknes, morocco<sup>5</sup>Professor and plastic surgeon, Mohammed VI University Hospital Marrakech, Morocco<sup>6</sup>Professor and plastic surgeon, Private Practinor in Marrakech, Morocco<sup>7</sup>Professor and plastic surgeon, Head master of plastic surgery department at Mohammed VI University Hospital In Marrakech, Morocco**\*Corresponding author**

Elatiqi Oumkeltoum

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**Abstract:** We report a rare case of a primary cutaneous chordoma in a 12 years old girl. 12 years old girl, without a history, presented since two months a subcutaneous lesion of the neck, removed and whose histological study spoke of a chordoma, lost side for 3 months, with notion of a multinodular neck tumor quickly evaluating. Removal of the entire tumor with wide resection margins and radiotherapy doses, the histological result speaks for primary chordoma with absence of other locations after a radiological assessment. Chordoma is a rare, locally aggressive tumor that rarely causes metastasis, sitting exceptionally at the level of the skin primitively, it is a tumor of the adult, poses difficulties of surgical excision due to its locations, good prognosis in case of skin localization, in our patient and with respect for therapeutic modalities (surgery and radiotherapy) the prognosis in our patient was very reserved, she died a year later.

**Keywords:** primary cutaneous chordoma, neck, chordoma, tumor, radiotherapy.

**INTRODUCTION**

Chordoma is a rare malignant tumor representing 4% of bone tumors, it is a dysembryoplastic tumor that derives from the remains of the fetal notochord, it is a cancer with slow local malignancy, with tendency to invasion of neighboring tissues, giving rarely metastases develops mainly in men aged between 50 and 60 but can occur at any age, seat preferentially in the sacro-coccygian (50%) and spheno-occipital region (35%) the axial location along the spine is found in 15% of cases, other exceptional sites have been reported: nasopharynx, orbit, frontal sinus, occipital scale, skin.

**OBSERVATION**

We report a case of cutaneous chordoma of cervical seat in a 12-year-old girl. H.E 12 years old, with no pathological antecedents, presents in June 2012 a nodular lesion of 2 cm in diameter next to the neck, an excisional biopsy with sending the product to the pathological study was performed. The anatomopathological diagnosis is a chordoma after an immunohistochemical study.

4months later, local recurrence with multiple large nodular lesions, subcutaneous fast growth, fixed with the superficial plane and mobile with the deep plane, a telangiectatic and inflammatory skin (figure1). A CT scan was requested: : multilobulated paravertebral mass projecting from the occiput to C5 (image2) well limited 13/7 cm diameter calcified taking the contrast product invading the left trapezius muscle without bone lysis (figure3).



Fig-1: Preoperative appearance of the tumor



Fig-2: Projection of the tumor From the occiput to C5

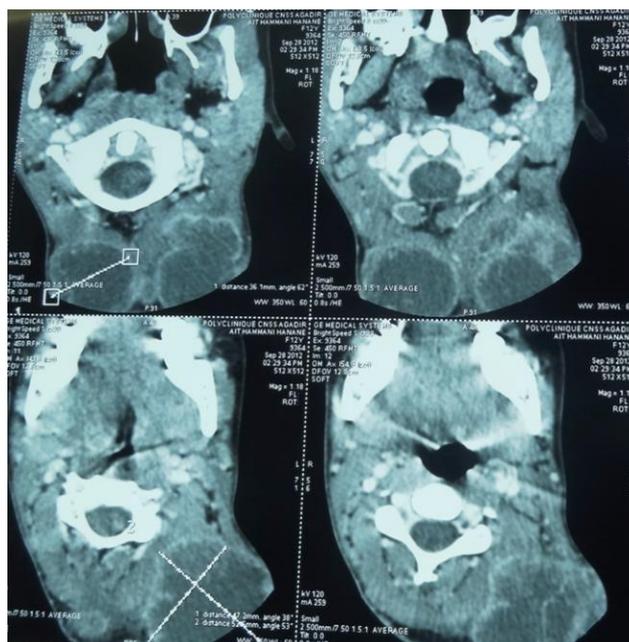


Fig-3: CT appearance of the tumor: lobulated appearance with calcifications

Patient sent to chemoradiotherapy with four chemotherapy sessions without any significant improvement. A cerebral CT scan was requested looking for an initial tumor and returned normal patient admitted to the neurosurgery department and transferred to the plastic surgery department in the absence of spinal or cerebral involvement. During hospitalization in neurosurgery, the tumor became fistulized with the notion of gelatinous fluid.

Upon admission to the plastic surgery department, it was clinically a nodular, fistulized, well-

limited tumor on telangiectatic and inflammatory skin 25 cm long / 12 cm width, occupying the whole neck and preventing the patient from raising hier head with no neurological deficit.

The first surgical procedure consisted of an excision of the tumor that invaded the left trapezius muscle which was partially removed (figure4) with sending of the piece to the anatomopathological study (figure 5).



**Fig-4: Complete excision in a single block of the tumor**



**Fig-5: surgical piece sent to the anatomopathological study**

The anatomopathological diagnosis came back similar to the first with the notion of malignant chordoma with deep grazing and superior tumor boundaries, on the rest the margins ranged between 1.5 and 2 cm. A second surgical procedure was performed

with resection margins (2 cm) deep and superior with a thin skin graft, and sending the removed product to the pathological study whose answer did not speak about a tumor residue.



**Fig-6: Represe of margins of exeresis with thin skin graft**

After complete healing and in collaboration with the radiotherapy department, the patient was sent for radiation treatment with high doses, with regular

follow-up every month to watch for a possible recurrence (figure 6).



**Fig-6: Appearance after complete healing and after radiotherapy after 4 months of evolution**

An extension assessment was performed with a cerebral CT, a thoraco-abdominopelvic CT completed by an abdominopelvic ultrasound not objectifying any anomaly.

After 8 months of follow-up without recurrence, the patient was lost to follow-up, and after one year she probably died from cerebral metastases because she had problems with consciousness according to her family

## DISCUSSION

Chordoma is a tumor that was first described by Ribbert in 1894, rare, represents 1 to 4% of malignant bone tumors and 1% of intracranial tumors, it is a tumor derived from vestiges of the fetal notochord [1] which are present in the nucleus pulposus of the intervertebral discs, vertebral bodies, sacrum, basilar process of the occipital and sphenoid[1]. Observed especially in men between 50 and 60 years old [2,3], in children it is exceptional and generally reaches the base of skull and the cervical spine. The frequency of localization of affected areas is subject to controversy [1,4], it is identical between the sacro-coccygeal, intracranial and cervicothoracolumbar spine according to McMaster *et al.* [8], the sacrococcygeal localization predominates in some authors 50% [7] while it is the intracranial localization that prevails in Meecherson *et al.* [8]. the clinical manifestations are very variable but the pain is almost constant, the insidious and latent nature of the tumor is at the origin of the delay of diagnosis.

It is a tumor with local malignancy rarely giving lymphatic or hematogenous metastases 5 to 60% [3], local recurrences are very frequent which explains the aggressiveness of the surgical procedure which must remove the whole tumor in monobloc with the widest margins of safety at least 1 cm, in some locations complete resection is not possible, therefore radiotherapy is indicated which should be performed in

association with surgery and never alone it is a high dose radiotherapy of at least 40 grays that can reach up to 80 grays (sarcum) for 7 to 8 weeks[2,5]. In metastatic forms, chemotherapy has been proposed and has given good results (disappearance of intracerebral metastases) [1,6].

The prognosis of chordoma depends on the size of the tumor and the possibility of radical excision reducing the risk of recurrence that can occur from the first month to 14 years, the survival rate is estimated at 5 years in 50% of cases, at 10 years in 25% and at 20 years in 5% of cases [2, 3].

## CONCLUSION

Cutaneous chordoma in children is a very rare tumor with evolutionary character very insidious, Radical surgical treatment combined with high-dose of radiotherapy can predict the occurrence of local recurrence and metastases that may occur in the short or long term, which explains the importance of strict clinical and paraclinical monitoring of patients.

## REFERENCES

1. Champeaux K, San-Galli F, Eimer S, Liguoro D. Métastase radicaire d'un chordome secondaire à une dissémination dans le liquide cébrospinal. Neurochirurgie. 2008 Feb 1;54(1):41-5.
2. Maaquili R, Moustaine M R, chordome sacré (à propos d'un cas), Rev. Maroc. Chir. Orthop. Traumatol. 2002, 16 : 63-66
3. Van Akkooi ACJ, Van Geel AN, caps report extra axial chordoma, The Journal of Bone and Joint Surgery. 1232-1234
4. Adam I. Rubin MD, Chordoma cutis J. Am acad dermatol. 52, (5): 105-108.
5. Jallo J, Nathan D, Bierbrauer K, Farber E. Chordoma: A case report. Surg Neurol 1997;48:46-8.

6. Jeantot C, Vinh Ts. Chordome sacré : revue de onze cas traités chirurgicalement, Rev. Chirorthop réparatrice. 2000, Nov, 86 (7) : 684-93.
7. Markwalder TM, Zimmerman A. Intracerebral ciliated epithelial cyst. Surgical neurology. 1979 Mar;11(3):195.
8. Adam R, McMaster P, O'Grady JG, Castaing D, Klempnauer JL, Jamieson N, Neuhaus P, Lerut J, Salizzoni M, Pollard S, Muhlbacher F. Evolution of liver transplantation in Europe: report of the European Liver Transplant Registry. Liver transplantation. 2003 Dec 1;9(12):1231-43.