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Case report

# Perineal chordoma cutis, a rare localization for a rare pathology: a case report and consideration of the literature

**Chennoufi Mehdi[1,&], Mhanna Tarik[1], Irzi Mohamed[1], Jdaini Ahmed[1], Boukhannous Ibrahim[1], Barki Ali[1]**

[1]Department of Urology, Mohammed IV University Medical Center, Mohammed the First University Oujda, Oujda, Morocco

[&]Corresponding author: Chennoufi Mehdi, Department of Urology, Mohammed IV University Medical Center, Mohammed the First University Oujda, Oujda, Morocco

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**Abstract**

The chordoma is a very rare notochordal tumor, the cutaneous localization (chordoma cutis) is even rarer and generally follows a local invasion of the integument even a metastasis. The clinical presentation is non-specific and the diagnosis is often late. It is confirmed by the immunohistochemical characteristics of the tumor. Total surgical excision is necessary in order to avoid the local recurrence which represents a frequent character of this tumor. We report the case of a young patient with perineal chordoma evolving from childhood. To our knowledge, our patient represents the first case of chordoma cutis of the perineum without a history of chordoma.

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## Introduction

The chordoma is a very rare malignant bone tumor, which develops from embryonic vestiges of the notochord. The evolution is slow but with great potential for local and metastatic aggressiveness [1]. The chordoma has three main locations: the sacrum, the cervical spine and the base of the skull. Extra axial chordomas, formerly known as parachordomas, are even rarer. These are soft tissue tumors that usually occur in the extremities, the lower part being more common than the upper part [2]. Extra axial chordomas have been included in the 2002 classification of the World Health Organization (WHO) of soft tissue tumors in the category of "tumors of uncertain differentiation" with mixed tumors and myoepithelioma [2]. The term chordoma cutis is used in the literature to designate the cutaneous localization of chordoma following the local invasion of the integument or metastatic. In this work, we describe a case of cutis chordoma in the perineal region without invasion or metastasis of a previous chordoma.

## Patient and observation

A 38-year-old male patient referred to the urology clinic for perineal mass evolving from childhood with a rapid increase in size. The patient was circumcised at the age of 3 years with no other medical history and no family history. The mass was pedunculated, mole spanking 72x54mm (Figure 1). The physical examination of the patient was unremarkable. The ultrasound objectified the vascularized tissue nature on Doppler. Computed tomography was made to determine the locoregional extension, returns negative. A resection in mass was practiced under spinal anesthesia, with surgical margins passing in healthy zone (Figure 2, Figure 3, Figure 4, Figure 5). The anatomopathological examination with immunolabelling with PS 100 found a tumor proliferation physaliphores rich in myxoid and cartilaginous range not ossified with undifferentiated cells at the periphery arranged in pseudo-papillary structures, associated with a fibro-myxoid tumor stroma and a non-ulcerated cutaneous plane. The immunohistochemical profile objectified the expression of pancytokeratins AE1/AE3 compatible with a chordoma. No signs of recurrence or metastasis after 3 months of follow-up (Figure 6).

## Discussion

In 1857 Virchow *et al*. described the chordoma for the first time [3]. The chordoma is defined as a rare malignant bone tumor that develops from the remains of the notochord [1]. The notochord is a fundamental structure for embryological development: it defines the primordial longitudinal axis of the embryo indicating the location of vertebral bodies. It also plays the role of inducing the ectoblast in the differentiation of the neural plate [4]. The notochord usually regresses during the $10^{th}$ week of gestation. The notochordal remains which persist in the pulpy nucleus of the cartilaginous discs could explain the anatomical predilection of the chordoma in the axial skeleton, the clivus, and the sacral regions. The chordoma represents approximately 1 to 4% of all bone malignancies [5]. The origin of the chordoma remains controversial. It should be distinguished from benign notochordal cell tumor (BNCT) of the spine. However, research has suggested that the chordoma may originate from BNCT, due to the common anatomical origin. Chordoma is a rare tumor that occurs in the last decades of life, especially adults over 40 years of age. Its incidence is 0.5 per million in the European population [6]. The prevalence of primary chordoma is 1 to 8% of all primary malignant bone tumors, and 20% of all bone tumors of the spine [4]. Chordoma is more common in the white population, with 0.08% in white subjects, 0.02% in black subjects and 0.07% in others (mainly Asian) [7]. The location outside the axial skeleton of the chordoma is even rarer than axial chordomas. They represent 6% of all chordomas. Historically

called parachordomas, peripheral chordomas or extra-axial chordomas, are more difficult to diagnose. These tumors have a histological similarity to axial chordomas and are also immunohistochemically similar to mixed tumors and myoepithelioma [8]. Skin lesions are exceptional. Generally, they follow the local invasion of the integumen but can also be metastatic. The term chordoma cutis is used for both situations [4]. Twenty six cases of chordoma cutis have been published in the literature since 1993 [1,3,4,9,10]. In a series of 207 chordomas, only 9% (19 cases) presented a cutaneous localization: 12 local recurrences, 6 extensions to the skin and a metastasis. Invasion of the skin is more common in the sacrococcygeal region [9]. Skin metastases of chordomas have been described on the scalp, face, trunk and upper and lower limbs [10,11]. To our knowledge, our case represents the first localization of chordoma in the perineal region, without pre-existing chordoma. The clinical diagnosis of cutaneous chordomas is difficult or even impossible when the lesion is outside the usual areas and/or if the patient has no suggestive history. Indeed, the dermatological aspect is aspecific and the chordoma can appear as a firm, bluish, domed, cystic, erythematous, translucent, flesh-colored or even reddish-brown nodule [9].

The clinical signs of chordoma cutis are often late, due to the long and insidious nature of the course as well as the ambiguity of the symptoms [12]. The chordoma is only confirmed when the tumor volume is large, explaining a fairly long diagnostic delay, between 12 and 24 months on average [13]. The diagnosis is essentially based on radiological means (CT and MRI) and the diagnosis of certainty is based on histological examination of the operating room or biopsy [14]. The pathology study confirms the embryonic origin. Macroscopically, the chordoma cutis appears as a very limited dermo-hypodermic mass of pale color with a mucoid appearance. Microscopically, the architecture is most often multi-nodular. The tumor cells are cohesive, organized in a network, cords, nests or clusters and are bathed in puddles of a basophilic substance, myxoid or even choroid in younger subjects. These cells are often of two types: small and ovoid or large. The physaliphor cells are characteristic and very suggestive. They are defined by a large size due to a cytoplasm vacuolated in "soap bubbles", these vacuoles pushing the nucleus towards the periphery. The oval or round nucleus is generally hyper-chromatic [15]. On immunohistochemical examination, the chordoma expresses Vimenyin and Cytokeratins as AE1/AE3, CK8/18 and CK19. EMA (epithelial membrane antigen) is expressed in almost all cases and 30 to 90% express the protein S100. The most useful marker for the diagnosis of chordoma is currently Brachyury: although it is also expressed by hemangioblastomas and certain testicular tumors, its nuclear expression has a sensitivity > 90% [16]. The treatment uses an aggressive surgical approach, in order to obtain a resection en bloc with the widest possible healthy surgical margins, otherwise, a recurrence may occur. Taking into account the extension of the lesion and/or the structures affected during the diagnosis, resection with free margins is not always possible. Without adequate margins, adjuvant radiation therapy should be administered to hope for an increase in hypothetical disease-free survival [17]. The recidivism rate is around 26% to 68%. The probability of metastasis reaches 20% to 60% of cases (skin, lung, lymphatic system). Radiotherapy in high doses allows analgesics, delays recurrences, but has no indication in curative treatment without association with surgery [19]. Chemotherapy does not modify the clinical course [5,18]. The chemoresistance of the chordoma is currently proven [19]. Prolonged postoperative monitoring of patients is necessary for these patients. Despite their slow growth, chordomas are locally aggressive and destructive. They tend to infiltrate locally into adjacent bones and soft tissue with local recurrence. This tumor can metastasize late in approximately 37% of cases [20,21]. The overall reported average survival is 6.29 years and the relative survival rates at 5, 10 and 20 years are 67.6%, 39.9% and 13.1% respectively [22].

## Conclusion

Extra axial chordoma, formerly known as parachordoma, is a rare and aggressive tumoral pathology of soft tissue. The cutaneous situation (cutis chordoma) is encountered in two cases: an extension of an underlying chordoma or that of skin metastasis of a chordoma. Our description relates to the first case of extra-axial chordoma without a history of axial chordoma with a location never described before: perineal situation. The rarity of this pathological entity makes the diagnosis difficult, based on the immunohistochemical study, especially when the patient has no particular history. Complete excision and a long-term follow-up seem judicious for patients with this rare pathology given the scarcity of data in the literature on recurrences and metastasis.

## Competing interests

The authors declare no competing interests.

## Authors' contributions

All the authors have read and agreed to the final manuscript.

## Figures

**Figure 1**: pedunculated and mole perineal mass measuring 72-54mm

**Figure 2**: preoperative photo of the chordoma

**Figure 3**: complete excision of the chordoma mass

**Figure 4**: stitches in reverse mercedes

**Figure 5**: the final resected tissue

**Figure 6**: the postoperative scar

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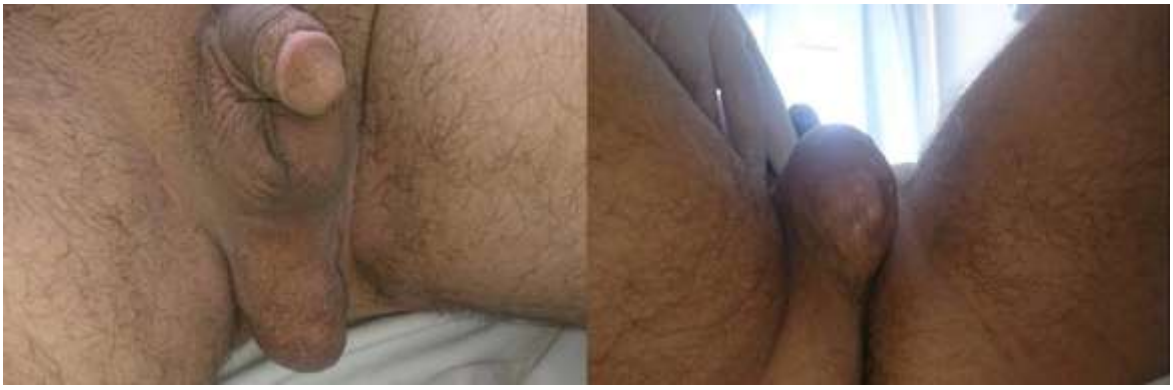

**Figure 1**: pedunculated and mole perineal mass measuring 72-54mm

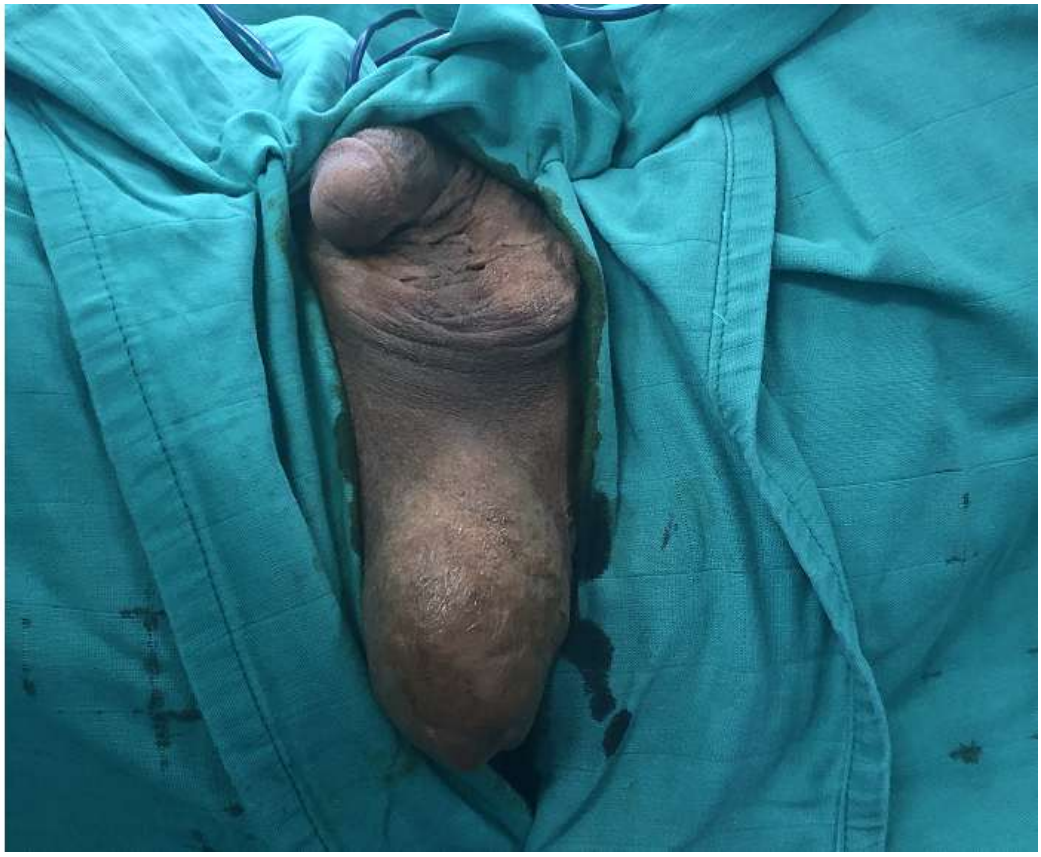

**Figure 2**: preoperative photo of the chordoma

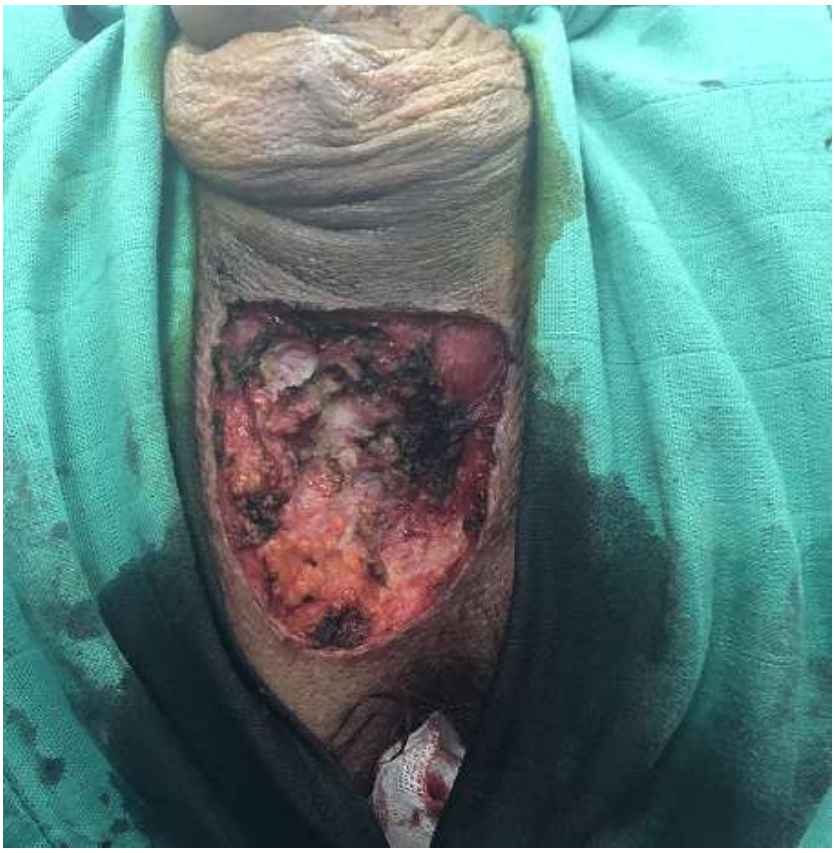

**Figure 3**: complete excision of the chordoma mass

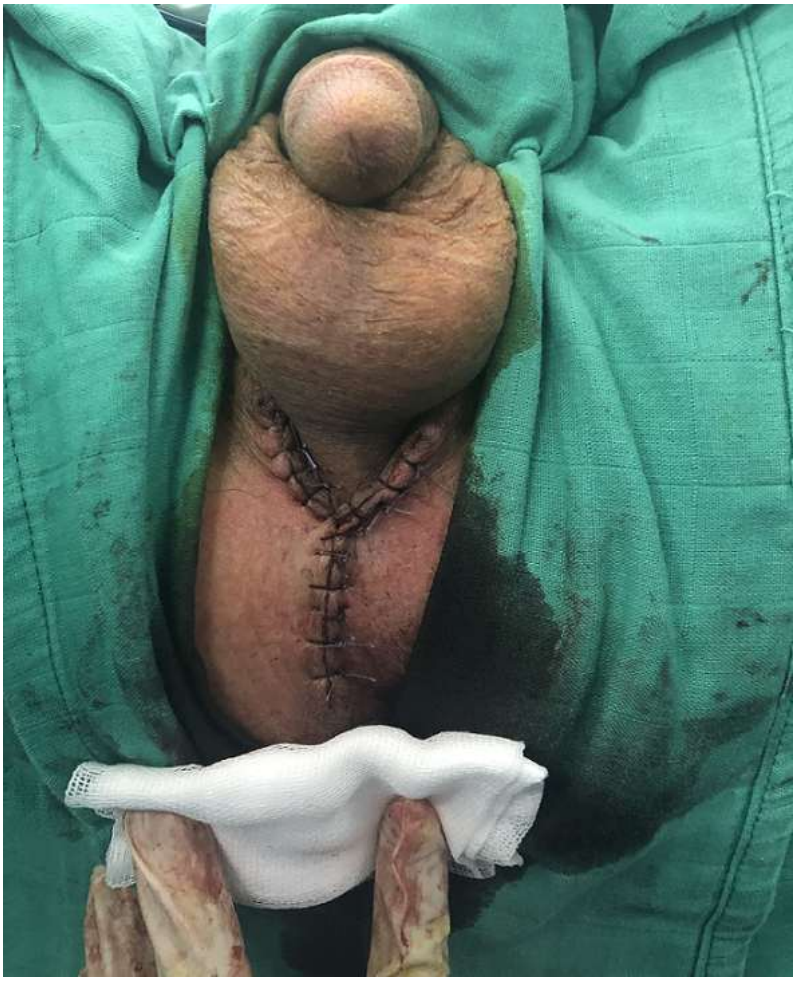

**Figure 4**: stitches in reverse mercedes

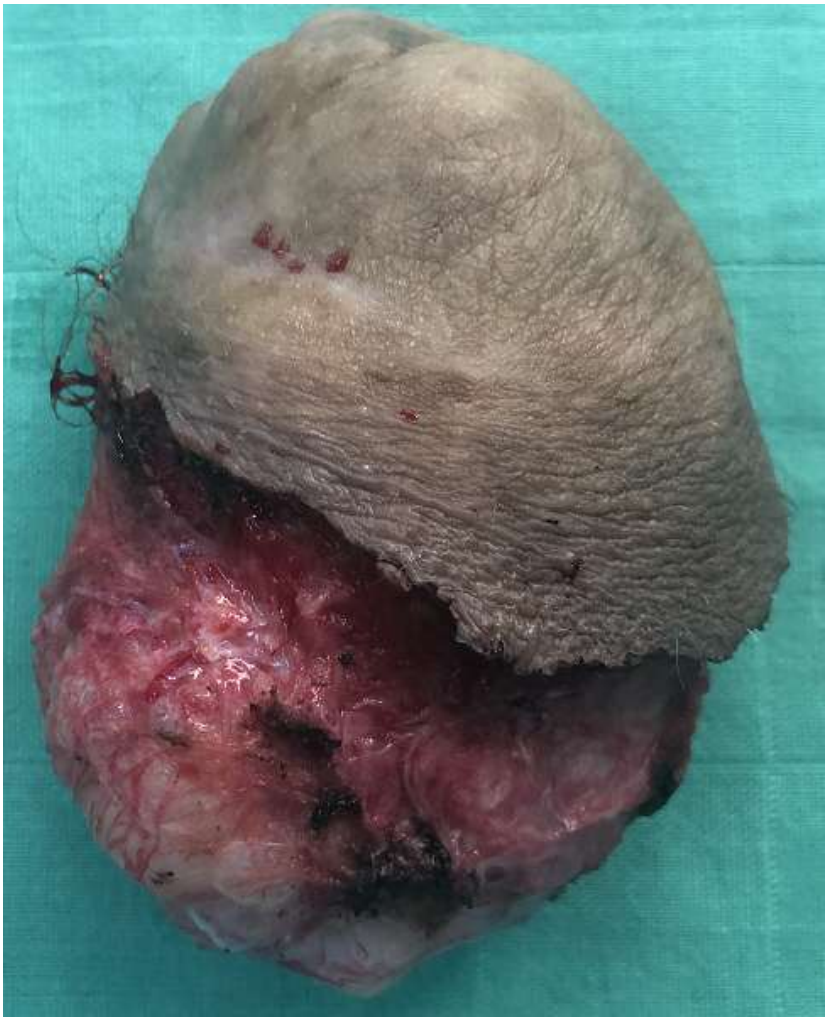

**Figure 5**: the final resected tissue

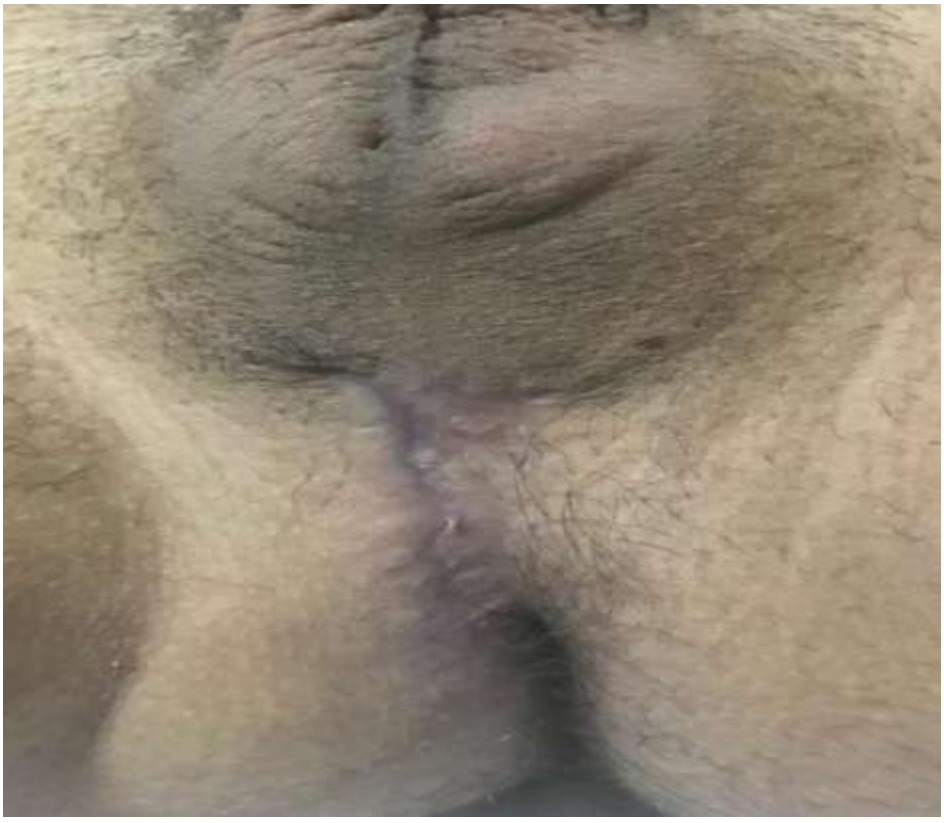

**Figure 6**: the postoperative scar