Highly differentiated sacrococcygeal teratoma.

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Citation

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Abstract

Sacrococcygeal teratoma is a well known entity and both its diagnosis and management have been thoroughfully described. Special mention deserves the highly differentiated teratoma as it presents complex organized tissular structures making it slightly different from the classic teratoma although it does not reach the defining characteristics of fetus-in-fetu.A female newborn without prenatal diagnosis presented sacro-coccygeal mass. X-ray image showed bone structures resembling tibia and fibula within the mass. She undertook surgery two days after delivery. Macroscopically, the mass showed besides the bones described on X ray image, cartilaginous structures, some loops of bowel and tissue resembling a finger. There were also some cystic areas. Highly differentiated teratoma also called fetiform teratoma seems to be a defined entity apart from the classical sacrococcygeal teratoma and the fetus-in-fetus. However differences regarding physiopathological and clinical aspects have not been yet completely described.

INTRODUCTION

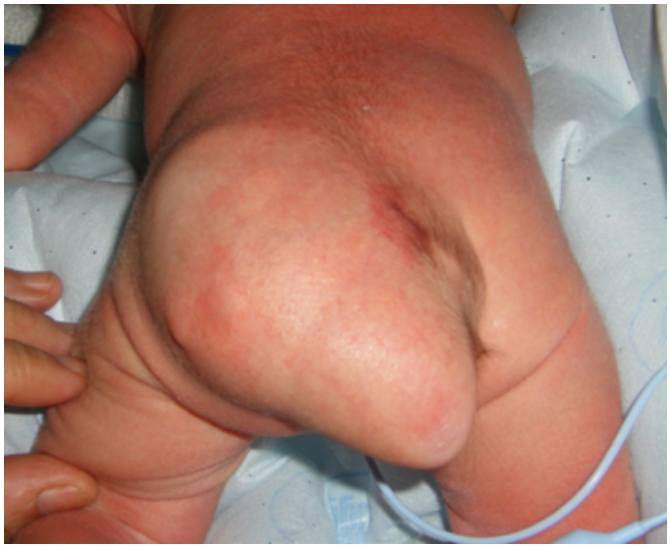
Sacrococcygeal teratoma is defined as a tumor emerging from proliferation of pluripotential cells, containing tissues from one or more germinal cell layers[1]. When differentiation of a teratoma includes complex tisular organization and organoid differentiation it is called fetiform teratoma or highly differenciated teratoma[2]. This is a less frequent condition and its physiopathology might be different from that of the classical teratoma although it seems to behave as a benign one. When the mass includes not only organoid differentiation but metamerization and spinal or vertebral axis as well it is defined as a fetus-in-fetu, an extremely rare condition with less than 100 cases published in literature[3].

CASE REPORT

A female newborn was delivered at 37 + 6 weeks of pregnancy, weighing 2770 g. It was a controlled pregnancy and she has no prenatal diagnosis. At first exploration she presented a sacrococcygeal mass 8 x 4,5 x 4,5 centimeters covered with apparently normal skin without external lesions (figure 1).

Figure 1

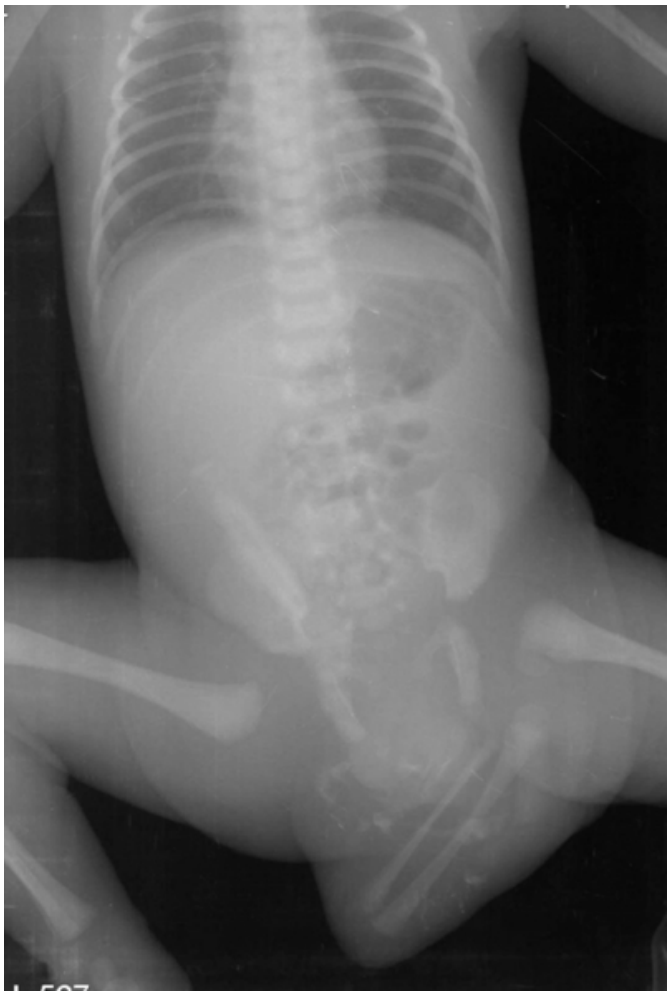
Figure 1. Sacrococcygeal mass



It was irregular and non symmetric, growing slightly to the right. X ray image showed a heterogenic mass with bones resembling tibia and fibula inside of it (figure 2).

Figure 2

Figure 2. X ray image



She undertook surgery at the second day. She was settled in prone position and a Chevron incision was made proximally, dissecting the mass distally until complete resection. She presented no complications during surgery and on early postoperative period and she was discharged ten days later. Anatomopatological exam revealed a heterogenic mass covered in skin with cartilaginous and fatty tissue, an intestinal loop, two long bones and an appendix resembling a finger. Alpha feto protein levels were 24,616 nanograms per milliliter (ng/ml) prior to surgery and decreased during follow up, being 20.05 ng/ml in last control, four months later. Beta human gonadotropine was within normal limits prior and after surgery. Nowadays she is asymptomatic and followed in our out patient service.

DISCUSSION

Sacrococcygeal teratoma is a congenital tumor present in 1 out of 40,000 newborns. It arises from caudal end of spine with different sizes and grades of protrusion. Most of them are benign, well differenciated and therefore their prognosis is good after surgical complete excision[1]. It is composed of one or more germinal cell layers originated fin pluripotential cells yet it lacks of complex tissular organization. According to the latest publications, the case we report can be considered as a highly differentiated teratoma[4]. This kind of tumor presents characteristics that make it slightly different from classical sacrococcygeal teratomas. It shows complex organoid differentiation, in our case, complete escheletal structures and bowel loops for instance, and it always behaves as a benign tumor, as we have reported too. However it does not reach the level of differentiation required to be considered as a fetus in fetu. This is an extremely rare condition defined by Willis as a mass containing vertebral axis and some other organs or limbs[5]. There are less than 100 cases reported in literature and its etiology is still unclear[3]. It seems to be accepted that a twin reabsorpted by the host is the origin of this kind of tumors, however some authors hypothesize that it could be the final stage in evolution of a highly differenciated teratoma and originate from pluripotential cells[6,7]. On the contrary, some other authors propose that both fetus in fetus and teratoma originates from aberrations of monozygotic twinning, therefore the pathology ranges from conjoined twins to fetus in fetus or fetiform teratoma depending on the timing of the embryologic alteration[8,9].

Further study would be needed in order to clarify the embryological origin of these tumors, define their characteristics and provide information that could help a better understanding and assessment of this pathology.

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