Radiology Case Reports 17 (2022) 2416–2423

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Case Report

Giant malignant sacrococcygeal germ cell tumor in a newborn: A rare case report☆

Carmela Brillantino[a], Maria Elena Errico[b], Rocco Minelli[c,], Giovanni Gaglione[d], Pietro Pirisi[d], Antonio Rossi[e], Biagio Francesco Menna[f], Marika Santarsiere[f], Mariateresa Rumolo[f], Eugenio Rossi[a]*

[a] *Department of Radiology - "AORN Santobono Pausilipon", Pediatric Hospital, Naples, Italy*
[b] *Department of Pathology - "AORN Santobono Pausilipon", Pediatric Hospital, Naples, Italy*
[c] *Department Life and Health "V. Tiberio", University of Molise, Francesco De Sanctis St 1, Campobasso, 86100, Italy*
[d] *Department of Surgery - "AORN Santobono Pausilipon", Pediatric Hospital, Naples, Italy*
[e] *Campus Bio-Medico University, Rome, Italy*
[f] *Department of Advanced Biomedical Sciences, University of Naples Federico II, Napoli, Italy*

ARTICLE INFO

Article history:
Received 21 March 2022
Accepted 30 March 2022

Keywords:
Newborn
Malignant sacrococcygeal germ cell tumor
Immature teratoma
Yolk sac tumor
Computed tomography
Magnetic resonance imaging

ABSTRACT

Malignant germ cell tumors constitute about 3%-4% of all neoplasms occurring before the age of 15. They arise in the ovaries, the testes, and in several other locations, including the lower back, the chest, the brain, and the abdomen. In infants and young children, the sacrococcygeal region is the most common site for extragonadal germ cell tumors, and teratomas account for the vast majority of sacrococcygeal germ cell tumors. Neonatal sacrococcygeal teratomas are usually benign and rarely they may contain a malignant component that is predominantly a yolk sac tumor. In this article, we describe a rare case of a male newborn with a giant sacrococcygeal mixed germ cell tumor composed of grade 3 immature teratoma and malignant yolk sac elements.

Introduction

Malignant germ cell tumors account for approximately 3%-4% of all neoplasms in children under 15 years [1,2]. They are a histologically heterogeneous group of tumors with a common origin in primordial germ cells, the embronic precursors of adult gametes. Malignant germ cell tumors occur predominantly in the gonads (ovaries or testes), but can also develop in extragonadal sites (sacrococcyx, mediastinum, intracranial region, retroperitoneum) due to the abnormal or arrested migration of germ cells precursors during embryogenesis [2–4]. In infants and young children, more than half of these neoplasms are extragonadal and sacrococcygeal teratoma (SCT) is the most common germ cell tumor found in newborns, occurring in approximately 1 in 35,000-40,000 live

☆ Competing Interest: The authors declare that they have no conflict of interest.

* *Corresponding author.*
E-mail address: rocco.minelli@libero.it (R. Minelli).

https://doi.org/10.1016/j.radcr.2022.03.107

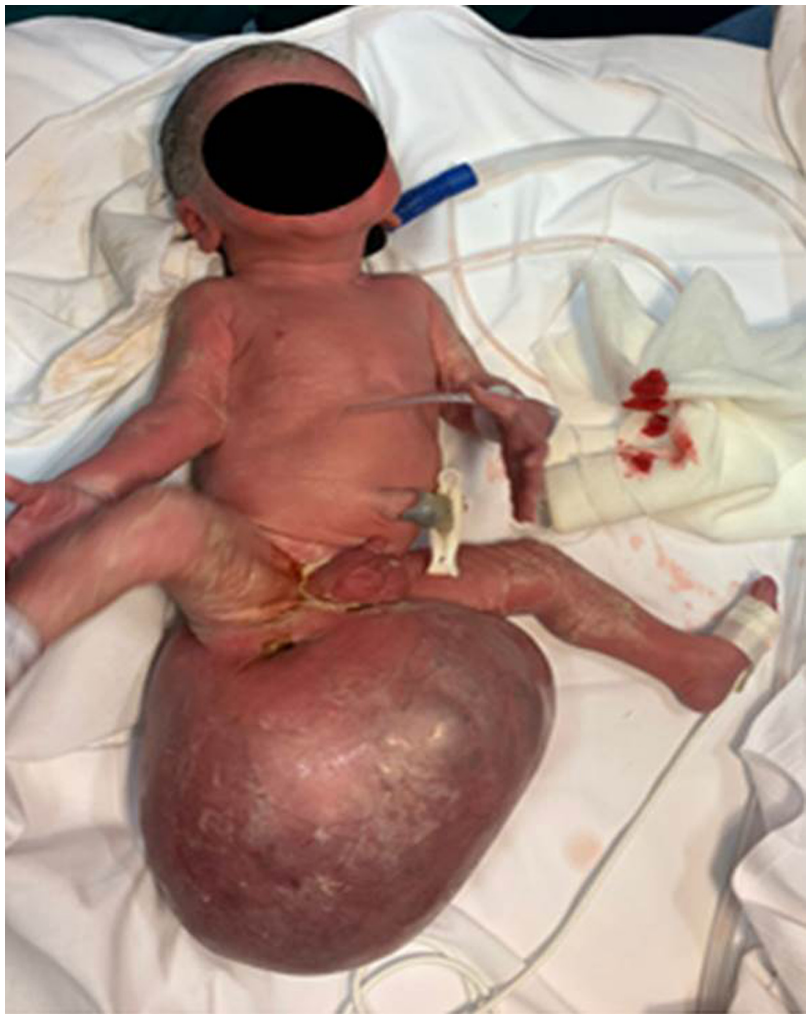

Fig. 1 – Clinical photograph of the baby a few minutes after the cesarean birth.

births with a female prevalence in a 3:1 to 4:1 ratio [1,2,4,5]. Immature and mature SCTs are usually benign and, in rare cases, they may be cancerous for the presence of malignant components, such as yolk sac tumor and less frequently embryonal carcinoma or choriocarcinoma [1,2,6,7]. Malignant SCTs represent about 12%-14% of sacrococcygeal germ cell tumors; they are not common in infants under 2 months and approximately 70% of SCTs are cancerous at 9 months of age [1,2,4–7]. In this paper, we report a rare case of a male newborn with a mixed malignant sacrococcygeal germ cell tumor containing immature teratoma and small foci of the yolk sac.

Case report

In August 2021, at our department of obstetrics and gynecology, a 4030 g baby boy was born at 37 weeks from 28 years old primiparous woman, after a pregnancy complicated by polyhydramnios. A routine second-trimester fetal ultrasound examination had found a mass located at the base of the spine, grown progressively in size. The family history was negative for birth defects and genetic disorders. An emergency cesarean section was performed due to decelerations on the cardiotocograph trace. The newborn had an Apgar score of 8 at 1 minute and 9 at 5 minutes. He was in no respiratory distress and had a giant tumor in the buttock area, with a maximum diameter of approximately 17 cm, a lobulated surface, and mixed solid and cystic consistency (Fig. 1). On the first day of life, the baby was transferred to our neonatal unit and subjected to laboratory and instrumental investigations. Complete blood count, liver and kidney function tests, electrolyte tests, blood glucose, urinalysis, and coagulation tests, were within normal limits, while serum alpha-fetoprotein (AFP) level was markedly elevated (172,000.0 IU/mL). The echocardiogram showed an ostium secundum atrial septal defect with a small left-to-right shunt. The abdominal ultrasound was unremarkable. Chest-abdomen-pelvis computed tomography (CT) and spinal magnetic resonance imaging (MRI) were then performed, both with intravenous (i.v.) administration of contrast medium (cm) and sedation. CT shows a large, predominantly exophytic mass extending from the coccygeal region; the lesion had an inhomogeneous structure due to the presence of solid components that appeared highly vascularized, and several fluid-filled areas with mild peripheral

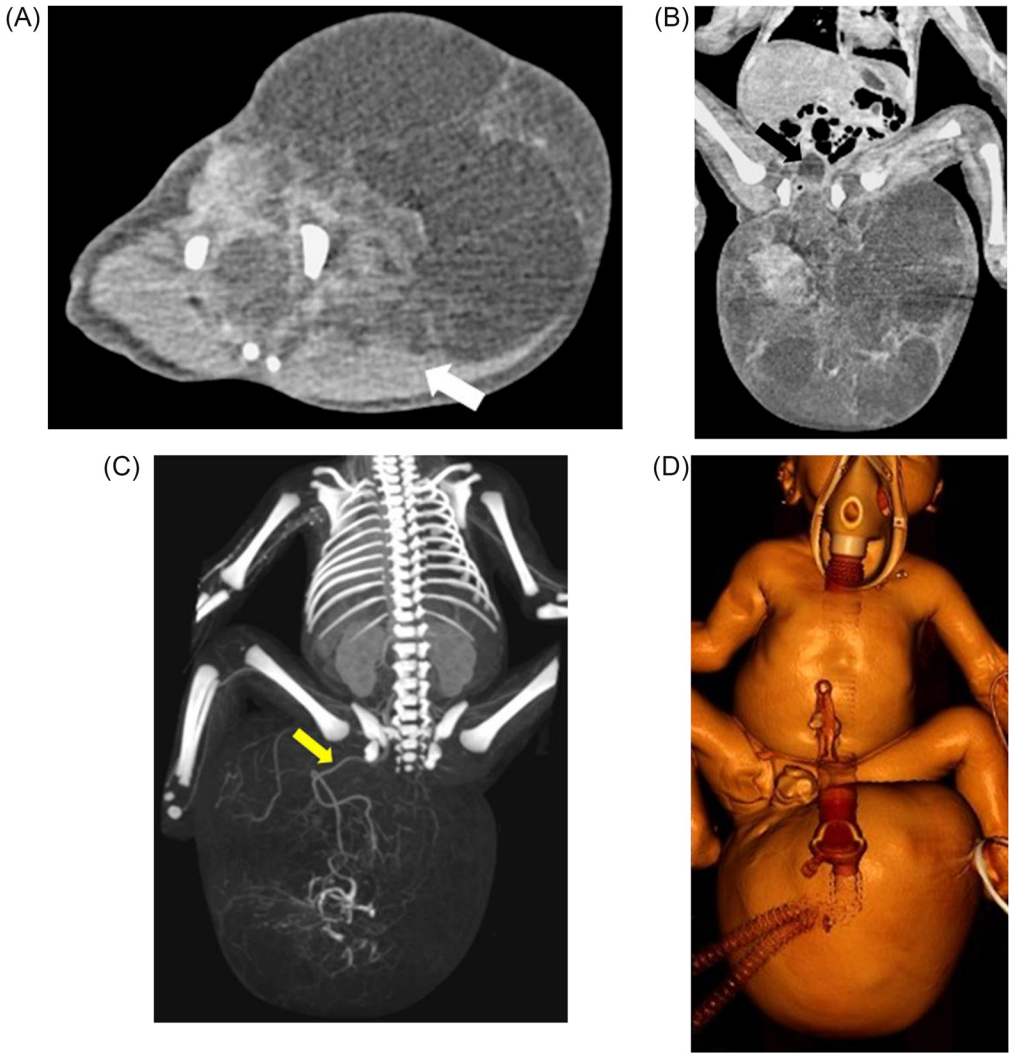

Fig. 2 – (A-B-C-D) Axial (A) and coronal (B) post-contrast CT images, highlight a voluminous heterogeneous enhancing mass occupying the lower pelvis with mostly external location. The neoplasm contains multiple areas of low attenuation consistent with cysts, separated by thin septa. It is attached to the left gluteus maximus muscle (white arrow in A) and displaces the bladder anteriorly and superiorly (black arrow in B). Right posterior oblique coronal MIP image (C) shows that median sacral artery, particularly apparent (yellow arrow), is the main blood supply of the tumor. Volume-rendered image of the baby (D).

rim enhancement. The tumor blood supply was implemented through a median sacral artery and vessels arising from the left internal iliac artery. The neoplasm caused compression of the rectum (including the anorectal junction) pushed to the right front, and of the bladder dislocated anteriorly and superiorly, without signs of infiltration of these organs and of anorectal/urinary obstruction. The bony component of the pelvis was undamaged and there was no evidence of secondaries in the chest or abdomen (Fig. 2). Spine MRI highlighted the giant complex mass, largely exterior, arising from the coccyx. The solid components of the tumor appeared isointense on T1- and T2-weighted images, with marked enhancement on T1 after gadolinium injection. On T2-weighted sequences, numerous cysts of varying sizes were noticeable within the mass, showing high signal intensity indicating simple fluid content. There was no local invasion, nor spinal canal involvement, and no other pathologic findings were detected (Fig. 3). The neoplasm was labeled as predominantly external SCT (type 1) and, at 4 days of life, the newborn underwent surgery. The chevron (inverted V) incision, with the newborn in the prone position under general anesthesia, was made, and the tumor was totally excised together with the coccyx, after ligation of its main blood supply, which was median sacral artery and peripheral branches of the left internal iliac artery in order to reduce operative blood loss. Reconstruction of the pelvic floor, buttock, and the preservation of the sphincter function was perfectly successful and there were no post-surgical complications. Grossly the large mass measured 17 × 11, 5 × 12 cm, and weighed 1, 5 kg; it was soft in consistency, with a bosselated external surface (Fig. 4). The cut surface showed a variegated appearance created by solid greyish white areas and cystic spaces. Microscopically, the

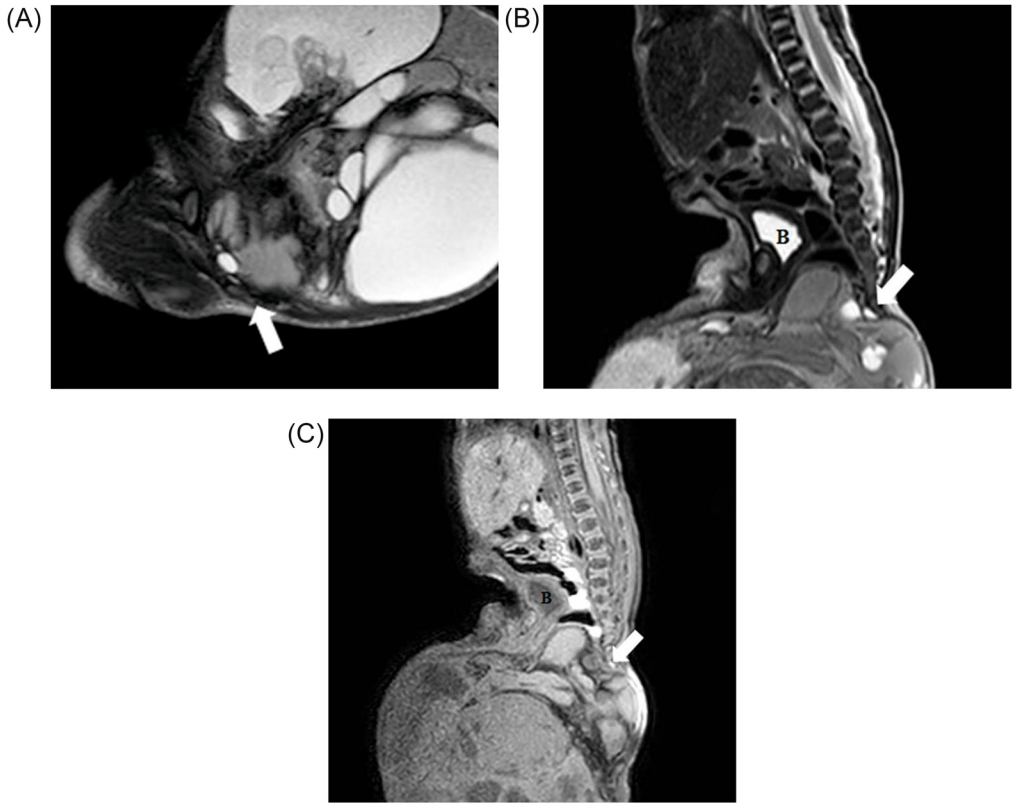

Fig. 3 – (A-B-C) Axial T2-weighted (A), sagittal T2-weighted (B), and sagittal T1-weighted post-gadolinium (C) images demonstrate a huge precoccygeal multicystic mass located postero-inferior to the bladder. A well-defined plane of cleavage between the tumor and the anterior surface of the last coccygeal vertebrae was not appreciable, but there was no invasion of the spinal canal (white arrows).

lesion, covered by skin, showed several cysts lined by squamous or cylindrical epithelium, mucus secreting glands, choroid plexus, fibroadipose, and neuroglial tissue, and hypercellular cartilage; moreover, immature mesenchyme with rhabdomyoblastic differentiation and areas of small undifferentiated cells, forming primitive neuroepithelial elements (>4 Low Power Fields), were present (Fig. 5). In this background foci of embryonal glands, lined by columnar cells and endodermal cysts (positive for panCK, Alpha-Fetoprotein, and SALL4) were observed (Fig. 6). The final diagnosis was grade 3 immature teratoma with foci of yolk sac tumor. No neoplasm was found in the coccyx fragments. During a postoperative hospital stay, chest X-ray didn't show alterations, AFP levels have decreased rapidly, and serum thyroid function test revealed congenital hypothyroidism (TSH 102.300 μIU/mL and free T4 0.56 ng/dL); ultrasound of the neck showed a thyroid normal in position, size, shape, and echotexture, and levothyroxine treatment was given. The little patient was not subjected to adjuvant chemotherapy and was discharged 45 days after birth in a good general condition, with AFP level declining to 339.1 IU/mL, improvement of thyroid function, negative abdominal ultrasound, and MRI of the brain, spine, and pelvis which did not detect pathological findings. The baby is currently undergoing careful follow-up at our hospital that comprises physical, instrumental, and laboratory examinations.

Discussion

SCTs are extragonadal germ cell tumors, the most common neoplasms in newborns, and are found mainly in females; frequently the fetal ultrasound, in addition to identify the mass, diagnoses the polyhydramnios, with an increased risk of premature delivery, as happened in our case. The incidence of congenital anomalies associated with SCTs ranges from 5% to 43% and anorectal and genital malformations constitute the major part of these. We found an SCT in a newborn who also had an ostium secundum atrial septal defect and congenital hypothyroidism, but nothing similar appears to be reported in the literature [1,2,5–7]. SCTs are composed of different types of tissue derived from at least 2, or the 3 embryonic layers (ectoderm, endoderm, and mesoderm); depending on the tissues that are included, they are classified as mature (fully differentiated components such as fat, bone, cartilage, teeth, and hair), immature (embryonal elements or incompletely differentiated structures), and mixed malignant (mature and/or immature types, together with mostly foci of yolk sac tumor and less often of embryonal carcinoma or choriocarcinoma) [1,4–7]. SCTs in newborns are usually mature and immature, with benign clinical course, but are more likely to be cancerous in males, as in our case, or if surgical resection is delayed;

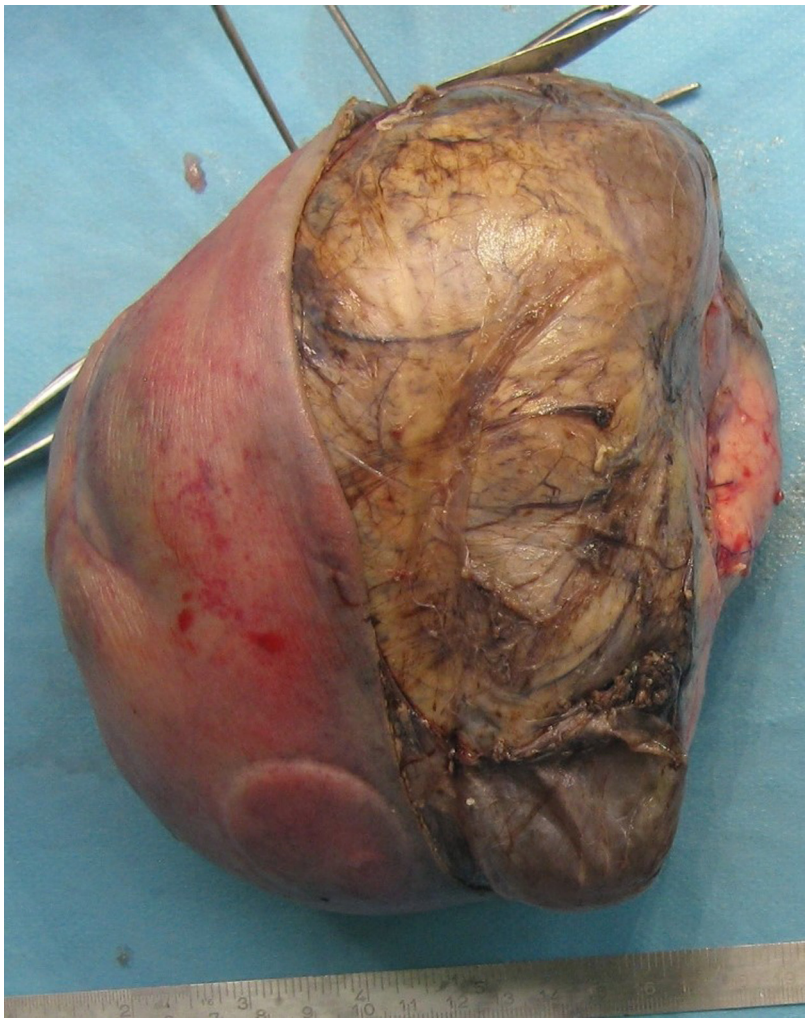

Fig. 4 – Gross appearance of the tumor.

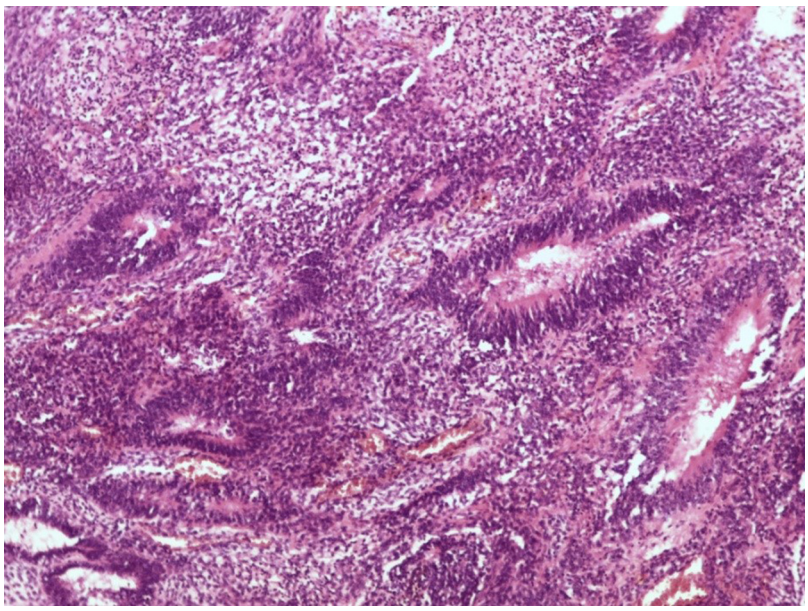

Fig. 5 – Tubules of primitive neuroepithelium in immature teratoma (H&E, 200x).

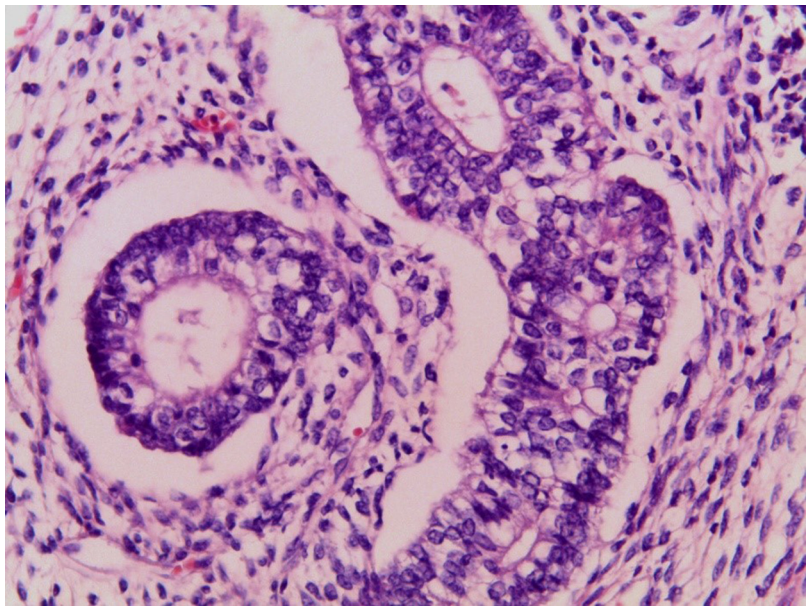

Fig. 6 – Yolk Sac Tumor. Embryonal glands lined by columnar cells with basal vacuoles (H&E, 400x).

moreover, the incidence of malignancy increases with age, being about 10% in the neonatal period and approaching almost 100% by the age of 3 years [5–9]. Although SCTs are classified according to the absence (grade 0) and the amount of immature tissue (grades 1, 2, 3) the histological grade of immaturity is not correlated with prognosis and only the presence of yolk sac tumor elements is predictive of malignant behavior. We found a grade 3 immature teratoma with yolk sac components, confirming, however, the known association between high-grade neoplasms and microscopic foci of malignancy [1,2,6,10]. Elevated serum AFP is considered the best indicator of yolk sac tumor, but this association must be interpreted carefully as the AFP is physiologically high in newborns because of fetal production and gradually declines to the normal adult range by 8 months after birth. Our little patient was considered to have elevated AFP levels for age consistent with the diagnosis and decreased quickly following surgery. After resection of SCT, AFP has a good predictive value for the presence of residual disease and its serial measurement is useful in the follow-up as, in absence of recurrence, it should fall and normalize within 6 to 12 months (9 months on average) [2,4–7,10–11]. Clinical presentation varies depending on the size and location of the SCTs. These neoplasms are usually asymptomatic, as in our case, but occasionally their growth within the pelvis can cause symptoms of obstruction of the rectum, colon, or bladder outlet, and/or lower limb dysfunction if the intradural spinal involvement is present [1,2,6,7,12]. The American Academy of Pediatrics Surgical Section classifies SCTs into 4 types based on the location, the ease of resection, and the malignant potential: type I (47%), predominantly external masses with a minimal presacral component, and carry the best prognosis; type II (35%), external tumors with a large intrapelvic portion; type III (10%), lesions apparent externally but predominantly pelvic and extending into the abdomen; and type IV (8%), presacral tumors only. The latter has the worst prognosis since, being devoid of external components, are difficult to diagnose, therefore, frequently malignant at the time of presentation as detected later than types I, II, and III, and sometimes less amenable to surgery. Contrary to what might have been expected, we found a type I SCT, which was cancerous [4–8,12]. Cross-sectional imaging studies such as CT and MRI play a central role in the management of SCT because, in addition to providing invaluable information that helps determine the diagnosis, define the type of mass by delineating its extension and relationship with the adjacent anatomical structures, so as to develop an appropriate surgical plan that enables complete resection of the neoplasm including coccygectomy [2,6,12–15]. CT and MRI are also very important in the evaluation of giant (>10 cm) hypervascular SCT, which is associated with a high risk of rupture and consequently severe bleeding during the operation unless the tumor is devascularized by ligation of the median sacral artery, which is considered its principal feeding vessel [5,7,8,16,17]. As in our case, the MRI is primarily used for evaluation of intraspinal invasion and for excluding the meningocele and myelomeningocele which are birth defects, grouped together under the term spina bifida cystica, looking like a sac filled with fluid leading out from the newborn's spine. Moreover, MRI is the modality of choice for the detection of neoplastic recurrence during follow-up, as it does not expose children to ionizing radiation and provides superior soft-tissue characterization to CT, exploiting the properties of magnetic fields to obtain images with high contrast resolution, acquired directly in 3 dimensions [6,7,9,10,13–15,18,19]. SCTs appear as complex masses, containing variable amounts of solid heterogeneous and cystic areas with or without septations, and imaging findings alone cannot differentiate between benign and malignant lesions. Generally, a large cystic component, calcification, and fat tissue are features of the benign SCT, while the malignant SCT is predominantly solid with fairly frequent hemorrhage

and necrosis. Our case, therefore, confirms that imaging cannot reliably predict the histopathologic diagnosis as the tumor contained numerous cysts with simple fluid content, was primarily external in location, and seemed benign; on the other hand, the destruction of the bones of the pelvis, invasion of surrounding organs or metastatic disease were not present [6,7,13–15,19]. Total excision of the mass including coccygectomy within the few days of life is the main treatment for malignant SCT. Currently, the risk of postoperative complications following excision of SCT is low, and constipation is the most frequently reported symptom [1,4–6,8]. There is no consensus on the use of adjuvant chemotherapy in newborns with completely resected immature teratoma containing malignant elements because very young age of the patients and acute and long-term side adverse effects of treatment. We adopted the "wait and watch" policy and chemotherapy will be initiated if there is evidence of reappearance of the disease based on rise in AFP and/or the presence of neoplastic tissue at MRI [1,20]. The recurrence rate of malignant teratoma is 0%-36% (mean, 18%) and the most important risk factors are incomplete resection of the neoplasm and/or coccyx, and/or tumor rupture/spillage during the cesarean delivery or surgical removal [4–8,10,12]. The probability of relapse of the disease is high in the first 3 years after surgery, but our little patient will undergo close follow-up for at least 5 years with frequent AFP level measurements alternated with diagnostic imaging examinations (abdominal ultrasound, chest x-ray, and pelvic/spinal MRI) [4,20].

Conclusion

Although rare, SCT is the most common tumor in the neonatal period and it is usually benign. We presented a rare case of a male newborn with malignant SCT that has a good overall prognosis. Management consists of complete excision of the neoplasm, including coccygectomy, with close follow-up for at least 5 years to detect recurrence. Imaging plays an essential role in the diagnosis, surgical planning, and follow-up.

Patient consent

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki declaration of 1975, and its late amendments. Additional informed consented was obtained from all patients for which identifying information is not included in this article.

Ethical considerations

Experiments on human subjects should be carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki). In the manuscript that informed consent was obtained for experimentation with human subjects. The privacy rights of human subjects must always be observed.

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