CASE REPORT

Neonatal sacrococcygeal teratoma with acute renal failure

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SUMMARY

Teratomas are germ cell tumours composed of multiple types of cells derived from more than a single germ cell layer. The most common site of an extragonadal teratoma is the sacrococcygeal region. We report a case of a 16-day-old female child with a large swelling in the sacrococcygeal region extending laterally into the buttocks with severely deranged renal functions. Ultrasonography and CT helped in making the diagnosis and, more importantly, to delineate the extent of the tumour and the involvement of adjacent organs and tissues: in our case, lower bilateral ureters. Imaging findings and clinical presentation led to the diagnosis of sacrococcygeal teratoma with renal failure.

BACKGROUND

Sacrococcygeal teratoma (SCT) is a rare tumour with a reported incidence of 1 in 35 000–40 000 live-births. However, it is the commonest solid organ tumour presenting in the neonatal period. Approximately 80% of affected infants are female. These are encapsulated tumours and the tumour originates during embryonic development owing to the failure of differentiation of primitive streak into the mesodermal, ectodermal and endodermal tissues, thus containing tissues from one or more germ cell layers.¹ Usually diagnosed at birth, the presentation varies from well-differentiated benign cystic lesions (approximately 70–80%) to malignant solid masses, incidence of malignancy increases with increasing age. A considerable variation in size is observed and it does not reflect the biological behaviour of the tumour. Prenatal ultrasonography (USG) allows the detection of tumour and its complications generally in the second trimester. Postnatal imaging has a role in establishing the extent of tumour and involvement of adjacent organs and tissues. We report this rare case of a large mature SCT with intrapelvic component and bilateral hydronephrosis in a neonate.

CASE PRESENTATION

A 16-day-old neonate from India presented to the outpatient department of our hospital with a large swelling in the sacrococcygeal region. The baby was reportedly born premature to a healthy 25-year-old mother after an uncomplicated gestation of 34 weeks through the normal vaginal route. The antenatal and perinatal period was uneventful. There was no history of any antenatal visit or USG of the mother. The initial examination revealed a cystic, non-mobile, non-tender mass approximately 7.5×6 cm in size in the right gluteal region

(figure 1). The overlying skin appeared normal with no evidence of any erythema, ulceration or active bleeding. Routine investigations were advised along with USG and CT abdomen for the characterisation of mass, to determine its extent and to aid in diagnosis.

INVESTIGATIONS

Laboratory investigations revealed markedly deranged renal function tests (blood urea nitrogen 130 mg% and serum creatine 7 mg%). USG of the swelling revealed mixed echogenicity with large anechoic cystic areas, scattered tiny hyperechoic areas and good vascular supply. An ill-defined mass extension in the pelvic region was also evident. For better anatomic delineation of the mass and determination of internal composition, non contrast enhanced CT (NCCT) of the abdomen and pelvis was advised to the patient, since a contrast-enhanced examination was contradicted, owing to deranged renal functions. The NCCT findings revealed a large well-defined, heterogeneous, dumbbell-shaped hypodense mass lesion with CT attenuation value of 20–30 HU with internal hyperdensities suggestive of calcification with intrapelvic, presacral and extrapelvic components (figure 2). The fat planes with the urinary bladder and rectum were well maintained with anterior displacement of the urinary bladder and anterolateral displacement of the rectum. Gross bilateral hydronephrosis was also present.

DIFFERENTIAL DIAGNOSIS

On the basis of clinical and radiological features, the diagnosis of mature SCT was considered. Other differentials were neurogenic tumours (neuroblastoma and ganglioneuroma), anterior sacral meningocele, sacral chordoma and rhabdomyosarcoma.

TREATMENT

Considering the urgency of the situation and poor general status of the premature baby, it was decided to perform a staged resection. Initially, surgical excision of the intrapelvic component of the tumour was performed to provide relief from urinary and bowel pressure symptoms. The transabdominal approach was used to remove the bulk of the tumour.

OUTCOME AND FOLLOW-UP

Histopathological specimen of the resected tumour showed the presence of elements from the three germ layers (figure 3). No immature cells or

To cite: Ahmad M, Arora M, Ullah E, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009304

Figure 1 Clinical photograph of the patient showing the swelling in the right gluteal region.

malignant features were seen, thus confirming the diagnosis of mature SCT.

All of the residual tumours as well as the coccyx were removed at a later operation after the improvement of the baby. She was discharged with advice to come in follow-up surgical outpatient clinic. There is no evidence of tumour recurrence till now at nearly 1 year of age. She had normal growth curve for her age with normal developmental milestones.

DISCUSSION

A teratoma refers to an encapsulated tumour with components resembling normal derivatives of all the three germ layers and usually arises as masses in the sacrococcygeal region.²⁻⁴ They are thought to arise from the totipotent cells from the Hensen's node at the anterior aspect of the distal sacrum and coccyx. Their predilection for this area is most likely due to the large number of totipotent cells usually found in the caudal region of

the embryo. Approximately 50–70% of SCTs are manifested within the first few days of life, and 80% are diagnosed before the age of 6 months.

Being encapsulated, teratomas are usually benign. Malignant transformation may occur, mainly into squamous cell carcinoma with increasing age; therefore, early diagnosis and surgical intervention are important. A mature teratoma is typically benign and is found more commonly in female infants, but immature teratomas are typically malignant and equally distributed among both sexes.

There are most often mixed solid/cystic, although purely cystic types can occur in ~15% of cases. Most SCTs manifest as presacral or pelvic masses. The masses also may extend into the abdomen. The commonly used system for classification (The American Academy of Pediatrics Surgical Section (AAPSS) classification) helps in grading the extent of SCT⁵:

- ▶ Type I: predominantly external masses with a small presacral component—50%.
- ▶ Type II: external masses with a significant intrapelvic component.
- ▶ Type III: external masses with a pelvic and abdominal component.
- ▶ Type IV: internal masses with an intrapelvic and abdominal location.

A special mention is to be made about type IV germ cell tumours, since their diagnosis is usually delayed, they are most likely to be malignant (approximately 38% of lesions).⁶ The diagnosis of a SCT is based mainly on histopathological examination. Imaging methods (USG, CT and MRI) performed alone or in combination are often pivotal for the detection of mass (especially type IV masses) to delineate their extension and assess soft tissue and organ involvement.^{7,8}

Prenatally, teratomas are usually diagnosed on the basis of obstetric USG in utero. They appear as a mixture of cystic and solid components. Recently, prenatal MRI has also been used in the imaging of antenatal fetal anomalies. In this case, the mother of the patient had not undergone any prenatal USG since she was illiterate and was not aware of the importance of prenatal USG in diagnosing neural tube defect in utero.

Figure 2 (A–D) The non-contrast-enhanced CT scan images showing a large, dumbbell-shaped, heterogeneous, hypodense lesion in the right pelvis extending to the ipsilateral extrapelvic region with internal hyperdense foci suggestive of calcification located peripherally.

Figure 3 The H&E stain sections ($\times 10$) of the resected tumour showing components resembling all the three germ layers suggestive of teratoma. (A) Stratified squamous lining with sebaceous glands and hair follicles, (B) sebaceous glands and hair follicles, (C) cyst lined by a flat epithelium and (D) mature cartilage and sebaceous glands.

The imaging features of these tumours depend on their contents.^{6–9} Benign teratomas contain predominantly mature tissues such as fat, fluid, soft tissues and calcification, while tumours containing large amounts of immature tissues are generally categorised as malignant.

Conventional imaging techniques have a very limited role in the evaluation of SCTs. Plain x-rays may reveal the areas of calcification within the mass. Barium contrast enemas may demonstrate the enlarged presacral space in these patients.

USG is the first line of investigation.⁹ Both USG of the mass and abdomen should be performed so as to characterise the nature of the mass, to look for any intrapelvic component, for other associated congenital abnormalities and also for potential complications that may develop such as bilateral hydronephrosis as in this case.

CT and MRI are the imaging methods of choice, their role being primarily to anatomically delineate the mass, and for visualising the involvement of neighbouring organs and tissues. In addition, certain imaging features can provide important clues that aide in diagnosis. MRI is also helpful in determining its connection with the vertebral column or its extension into the spinal cord. Although MRI has definitive advantages over CT, the limited availability of MRI, its high cost and patient cooperation are factors that favour the role of CT. In our case, non-contrast CT was performed (since the patient was with acute renal failure) and was very well able to fulfil the above-mentioned goals of imaging.

SCTs are known to cause urological complications including hydronephrosis, total urinary retention, hydrocele and even undescended testis.¹⁰ As in our patient, bilateral hydronephrosis can be present without discernable anatomical obstruction and appears to be due to poor bladder emptying or high intravesical pressures.¹⁰ Surgical intervention is usually required for prompt resolution of bilateral hydronephrosis or urinary retention.¹¹ Early surgical resection with complete excision of the coccyx is the intervention of choice.¹² However, our patient's poor general health necessitated the completion of excision in a staged manner.

Staged resection was advocated by Robertson *et al*¹³ by using the devascularisation technique through the ligation of vessels to

correct the hyperdynamic state, decreasing blood loss during surgery. However, it was admitted by them that this technique carried the theoretical risk of malignant transformation in the retained tumour if the histological grade is high and the residual tumour is left for long.

Learning points

- ▶ Sacrococcygeal teratoma must be considered as one of the differential diagnosis of swellings in the gluteal region.
- ▶ Sacrococcygeal teratoma can cause the obstruction of bilateral ureters leading to hydronephrosis and renal failure.
- ▶ In neonates with poor general condition, especially in premature neonates, the removal of the whole tumour may not be feasible in a single operation. In such cases a two-staged operation might be considered.
- ▶ Excision of coccyx must be performed to prevent recurrences.

Contributors All the authors had contributed to the work up of the patient including clinical evaluation, investigation and treatment planning. The manuscript has also been reviewed and approved by all authors.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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