RADIOLOGICAL CASE

CLINICS IN DIAGNOSTIC IMAGING (6)

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CASE REPORT

A nine-month-old Chinese girl was noticed to have a right buttock mass. There was no history of trauma, nor bowel or urinary symptoms. On examination, a large painless subcutaneous mass was palpable at the right sacrococcygeal region (Fig 1). It was oval-shaped, soft in consistency, and had a smooth outline. The overlying skin was normal. Rectal examination revealed a posteriorly located mass, the upper border of which was out of finger reach. Anal tone was normal and lower limb neurology was intact. The liver, kidneys and spleen were not palpable.

Serum α-fetoprotein level measured 39,550 ng/ml while β-human chorionic gonadotrophin (HCG) was less than 5 IU/L. Full blood count, renal and liver function tests, and urine catecholamines were normal. What do lumbosacral radiograph (Fig 2) and computerised tomography (CT) of the pelvis (Fig 3a, b) show? What is the diagnosis?

Fig 1 – Clinical photograph showing a large subcutaneous mass over the right buttock.

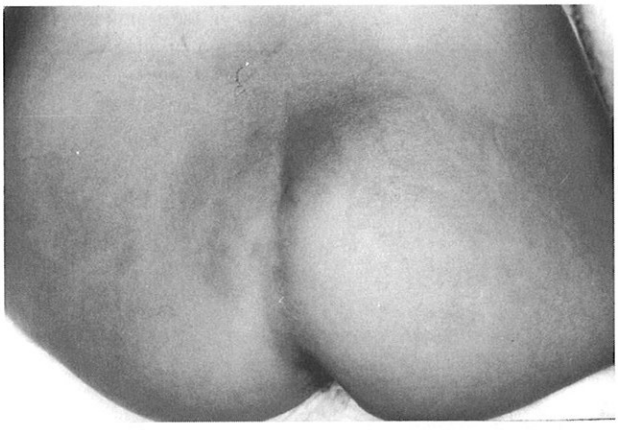

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Fig 2 – Lateral radiograph of the lumbosacral spine.

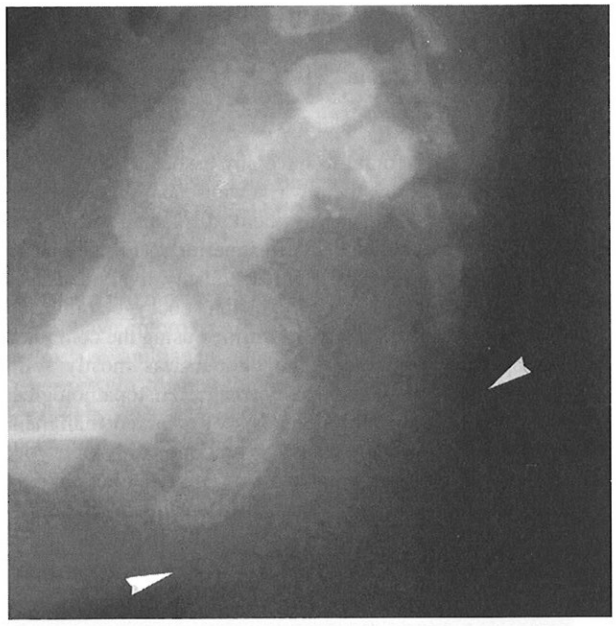
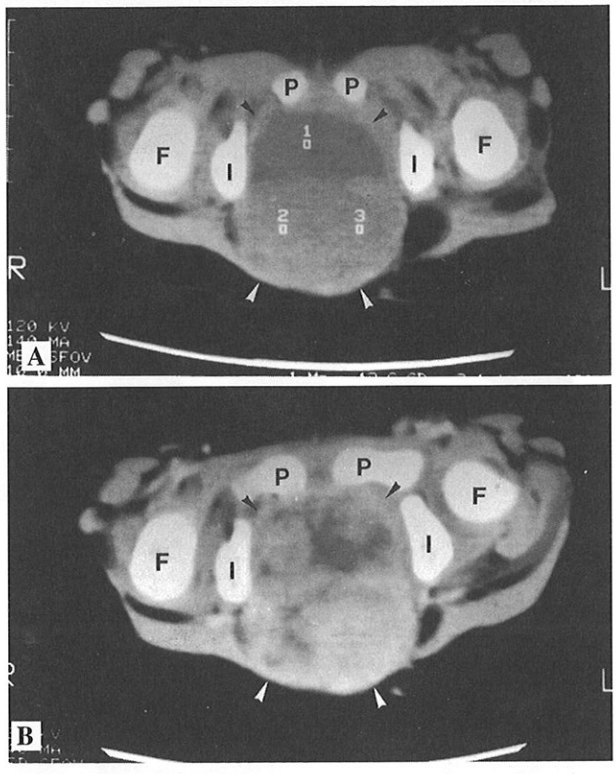


Fig 3 – (A) Non-enhanced and (B) enhanced axial CT scans of the lower pelvis at the trochanteric level.



Femur = F; Ischium = I; Pubis = P

IMAGE INTERPRETATION

Plain radiograph showed a suspicious soft tissue pelvic mass (arrowheads) distal and anterior to the tip of the coccyx (Fig 2). CT confirmed the presence of a heterogeneous solid mass (arrowheads) extending posteriorly from the lower pelvis. Plain CT demonstrated that the anterior part of this mass was of lower density (CT number 22 HU) compared to the posterior component (CT number 40 HU) (Fig 3a). The mass showed patchy enhancement (CT number measuring up to 90 HU) after intravenous contrast administration (Fig 3b). No calcification was detected. The exact extent and internal architecture of the mass was demonstrated in multiplanar fashion by magnetic resonance (MR) imaging (Fig 4).

DIAGNOSIS

Sacrococcygeal teratoma

CLINICAL COURSE

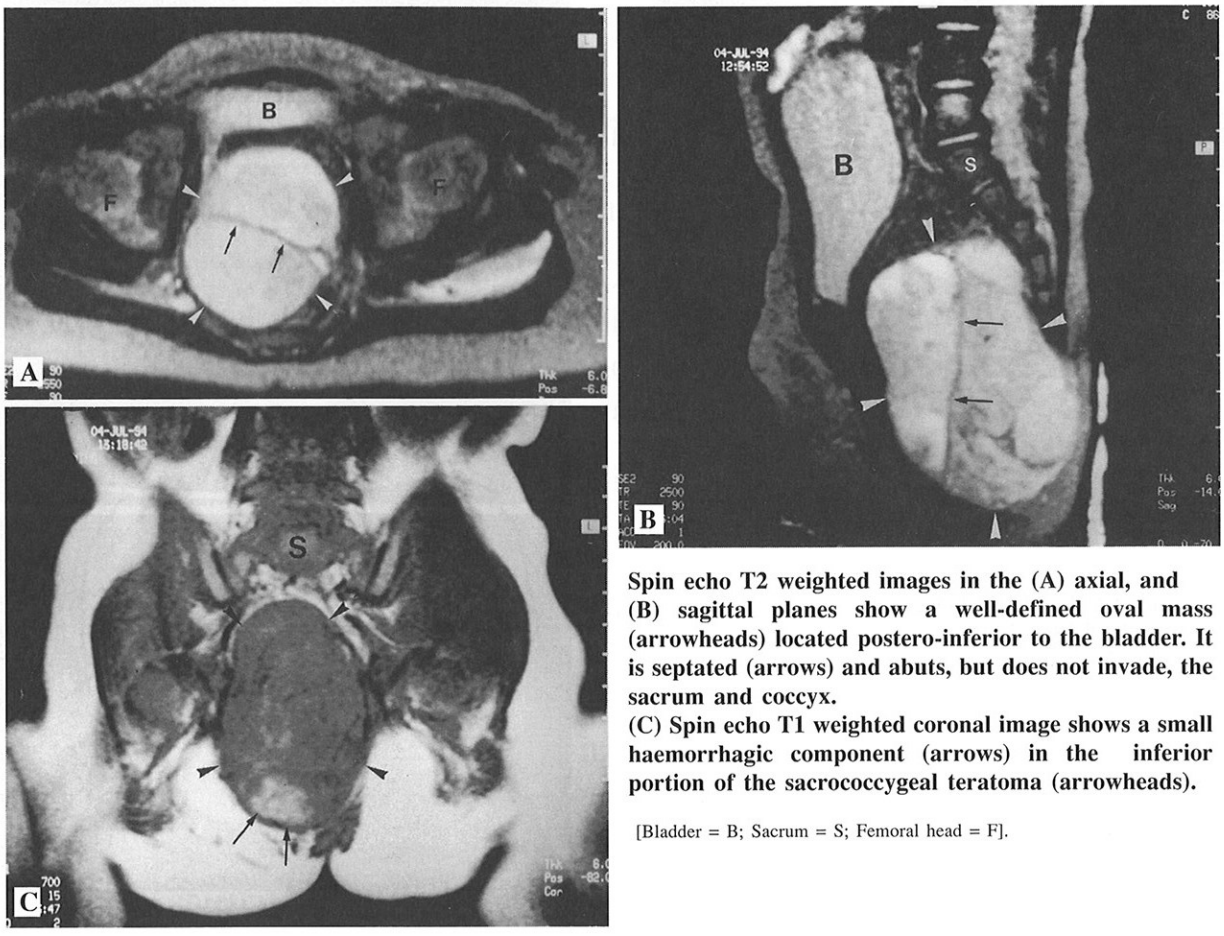
The patient was not found to have any associated congenital anomaly. At laparotomy, there was a well-encapsulated pelvic mass, measuring 7.5cm x 6cm x 5cm, displacing the rectum to the left and the bladder antero-superiorly (Fig 5). There was no demonstrable pelvic lymphadenopathy, liver metastasis or peritoneal seeding. Tumour excision, coccygectomy and Broviac catheter insertion were performed using the combined abdominoperineal approach. The tumour was mostly solid with areas of necrosis and haemorrhage. Histopathological examination confirmed the diagnosis of a mixed malignant sacrococcygeal teratoma; consisting predominantly of a yolk sac tumour, with minor components of embryonal carcinoma and teratomatous elements. The patient made a good post-operative recovery and was started on the multiagent chemotherapeutic JEB protocol [a cyclical regime consisting of JM8 (Carboplatin), Etoposide and Bleomycin]. Her serum α-fetoprotein levels fell progressively and dramatically to a normal level of 13 ng/ml after 2 months of chemotherapy. She remains well to date.

DISCUSSION

A teratoma is a true neoplasm comprised of multiple tissues of kinds foreign to the anatomical sites from which they arise. In children, the sacrococcygeal region is the commonest location of these tumours. The term sacrococcygeal teratoma encompasses a spectrum which includes the mature (benign), immature (containing embryonic elements), mixed malignant (yolk cell together with either mature or immature types) and pure endodermal sinus tumours[1-3]. Grading of sacrococcygeal teratoma according to the quantity of immature tissue present has been proposed[4].

Most sacrococcygeal teratomas present during the newborn period as large buttock masses. The tumour has a prevalence of one in 40,000 live births and predominantly occurs in girls (girl : boy ratio of 3-4 : 1)[1]. The clinical detectability of sacroccocygeal teratomas depends much on its anatomical location. Altman et al described a classification comprising four tumour categories, namely : Type I - predominantly external; Type II - external manifestation but have significant intrapelvic extension; Type III - apparent externally but predominantly pelvic with abdominal extension; Type IV - pre-sacral with no external manifestation[5]. Our patient's tumour was classified as Altman Type II. As expected, type IV tumours may not be detected till several

Fig 4 – MR scans of the pelvis.

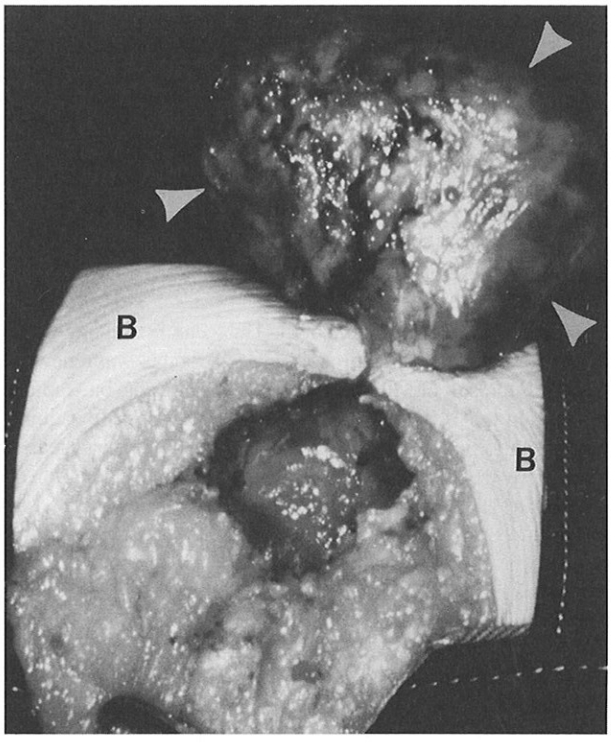


Spin echo T2 weighted images in the (A) axial, and (B) sagittal planes show a well-defined oval mass (arrowheads) located postero-inferior to the bladder. It is septated (arrows) and abuts, but does not invade, the sacrum and coccyx.

(C) Spin echo T1 weighted coronal image shows a small haemorrhagic component (arrows) in the inferior portion of the sacrococcygeal teratoma (arrowheads).

[Bladder = B; Sacrum = S; Femoral head = F].

Fig 5 - Operative photograph showing the sacrococcygeal teratoma (arrowheads), just prior to total excision. The patient is lying in a prone position. [Buttocks = B].



years after birth unless there are obstructive effects upon the urinary or intestinal systems.

The exophytic component of a sacrococcygeal tumour may be mistaken for a buttock tumour such as a lipoma or haemangioma, especially if a rectal examination is not performed to detect the pre-sacral component. Imaging using CT and MR helps define tumour extent and aids in differentiating sacrococcygeal teratomas from these tumours and other masses in this area such as meningocoeles, rectal duplication cysts and neuroblastomas[1,6].

Plain radiography is often able to demonstrate the soft tissue masses of types I and II sacrococcygeal teratomas, as well as in detecting associated congenital anomalies such as spinal dysraphism and sacral agenesis. The sacrum and/or coccyx may either be destroyed in malignant tumours or eroded in long-standing cases. Over half of the tumours have visible calcification or ossification. Ultrasonic appearances depend much on the composition of the sacrococcygeal teratoma, with the commonest pattern being a complex mass with equal solid and cystic components. Septation, fluid-fluid levels in cystic areas, and echogenic areas representing fat or calcification may be demonstrated[1]. Prenatal ultrasound is useful in the diagnosis of sacrococcygeal teratoma, monitoring its growth, and detection of associated complications such as hydronephrosis, polyhydramnios and foetal hydrops[7].

Both CT and MR imaging are the best modalities for defining the extent of the tumour. Sacrococcygeal tumours are typically midline in position, encapsulated and extend from tissues around the tip of the coccyx. On CT, they are frequently heterogeneous and enhancing; with fatty, calcified and cystic components readily identified using CT attenuation numbers. MR imaging, with its multiplanar capability, delineates the relationship with the adjacent soft tissue structures and spinal canal involvement particularly well. Metastases may occur in the lung, lymph nodes, liver, bones and brain. Imaging information thus provided facilitates pre-operative planning, though neither CT nor MR scans are reliable in predicting the histological characteristics of these tumours. Benign lesions tend to be largely cystic while malignant ones are likely to have substantial solid components[1,8].

Sacrococcygeal teratomas are optimally treated by complete surgical excision, combined with coccygectomy. In some cases, pelvic floor reconstruction may be necessary if the resection involves this structure. Patients with malignant tumours undergo chemotherapy, as well as surgery. Recurrences may however occur, even in benign sacroccocygeal teratomas. Benign tumours generally have an excellent prognosis while survival rates of the malignant forms have improved in recent years, due mainly to more aggressive multiagent chemotherapy[1,2,9]. The use of serum α-fetoprotein to monitor tumour response to treatment and for subsequent detection of recurrence has been recommended[9,10].

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ABSTRACT

A nine-month-old Chinese girl presented with a large buttock mass. Serum α-fetoprotein level was elevated. Plain radiographs, computerised tomography and magnetic resonance imaging demonstrated a solid heterogeneous presacral mass. A sacrococcygeal teratoma was excised using the abdominoperineal approach. The patient responded well to multiagent chemotherapy. The features of this tumour are discussed, with emphasis on the role of imaging.

Keywords: *germ cell neoplasm; foetus, neoplasms; neonate and children, neoplasms; sacrococcygeal teratoma; sacrum, neo plasm; yolk sac tumor.*