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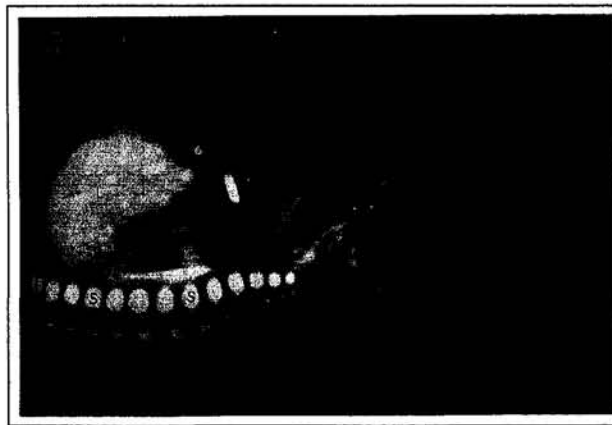
Clinical History:

A newborn baby girl of 23 weeks of gestation weighing 680 gm was found to have a large sacral mass. On palpation, the mass was firm in consistency. The anus and vulva were displaced anteriorly.

Figure 1: Clinical photograph of the premature baby taken shortly after birth. The sacral mass is arrowed



Figure 2: Direct sagittal CT scan of the lower trunk, taken through the midline. [S=spinal segment, L=liver]



Answer
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What is the diagnosis?

- a) Sacrococcygeal teratoma
- b) Sacral chordoma
- c) Myelomeningocele
- d) Myelocoele
- e) Lipomyelomeningocele

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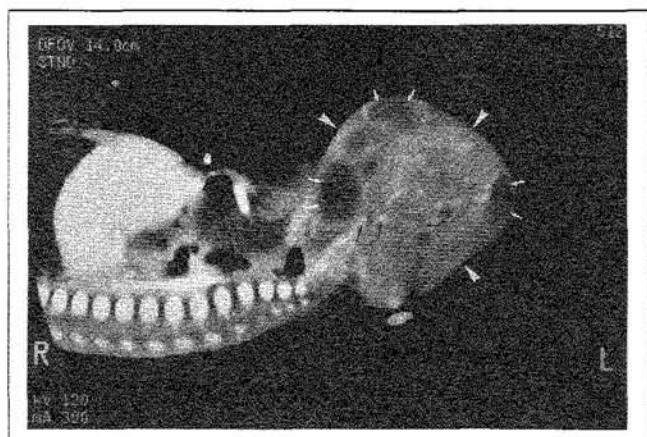
Answer:

- a) Type I sacrococcygeal teratoma

Radiological findings

The direct sagittal computed tomography (CT) scan showed a heterogeneous soft mass which arose below the level of rectum and extended distally from the coccyx. The mass was located in the midline and exhibited patchy contrast enhancement. In addition to the solid component, there were calcified and fatty areas (Figure 3). Findings were typical of a sacrococcygeal teratoma.

Figure 3: Figure is identical to Figure 2 with addition of arrows. The mass (arrowheads) is exophytic without intrapelvic extension. There are small hyperdense areas representing calcified foci (small black arrows), and larger hypodense areas representing the fatty components (small white arrows), within the mass



Discussion

A sacrococcygeal teratoma weighing 208 gm was successfully excised. The cut surfaces of the operative

specimen showed a variety of tissues (Figure 4). Microscopically, the tumour was found to composed of ectodermal (skin, squamous epithelium and neural tissue), mesodermal (adipose tissue and cartilage) and endodermal (respiratory and intestinal epithelium) tissues. This confirmed the pre-operative diagnosis of a teratoma, which is a complex neoplasm composed of tissues derived from all three dermal layers, and also explained the heterogeneous CT appearances. The patients made a good post-operative recovery and at the time of discharge from hospital, she weighed 2300 gm.

Figure 4: The resected surgical specimen of the teratoma shows a variety of different tissues which was confirmed histologically



The sacrococcygeal area is the most common site for teratoma in infancy with the second commonest site being the ovary.¹ Altman *et al* described the classification of sacrococcygeal teratoma in 1973.² CT is important for identifying the type of sacrococcygeal teratoma pre-operatively because the different types of tumour have different degrees of pelvic extension. The four types are: Type I, – predominantly external; Type II, – external manifestation but with significant intra-

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pelvic extension; Type III, – apparent externally but with predominantly intra-pelvic and intra-abdominal extension; Type IV, – pre-sacral with no external manifestation. Intra-pelvic extension, if present, requires an abdomino-perineal surgical approach for removal, which entails a much more major procedure. This is a particularly important consideration for our 680gm premature patient. The rectum and anus are sometimes markedly displaced as in our patient. Magnetic resonance imaging (MRI) of the pelvis, with its multiplanar capability, is especially suited in delineating the relationship of the tumour with the adjacent soft tissue structures and spinal canal.³

With improved quality of antenatal ultrasound, a large sacrococcygeal teratoma can be detected and managed at an earlier stage. High-output foetal cardiac failure due to vascular steal by arteriovenous shunts in the tumour and haemorrhage causing anaemia are major complications which may occur. Intrauterine endoscopic laser surgery has been reported to be a successful life saving procedure.⁴ Sometimes, devascularization and stage resection⁵ or cardiopulmonary bypass⁶ may be used for giant lesions.

Sacral chordoma

Chordomas are uncommon aggressive extradural lesions of the bony spine, which arise from primitive notochord remnants. 50% of the chordomas are located in the sacrococcygeal region, with 30-40% originating from the basisphenoid. They typically present in middle-aged adults. Plain radiographs show bony destruction and calcified areas. The solid paravertebral soft tissue mass, usually with associated septa, is better demonstrated on MRI than CT scans.⁷ In this case, the age, clinical presentation and imaging appearances exclude this diagnosis.

Myelomeningocele

Myelomeningocele is the most common significant type of spinal dysraphism. Due to failure of closure of the posterior neuropore at about 3 weeks of gestation,

there is a persistent neural tube defect which is manifested clinically as a midline back mass without skin covering. Patients have severe lower limb neurological deficits from birth, and often have neurogenic bladders. There is a high incidence of Arnold Chiari II malformation and other associated spinal anomalies. Radiographs show the large posterior vertebral defect and MRI is useful in demonstrating the herniated neural elements and cerebrospinal fluid (CSF), especially the presence of spinal cord tethering.⁸ In this case, the clinical and radiological features are against this diagnosis.

Myelocoele

Myelocoele refers to exposed neural tissue that is not skin-covered. Unlike myelomeningocele, the neural tissue has not been pushed posteriorly by CSF-filled meninges, therefore the patient does not have a back mass.⁸

Lipomyelomeningocele

Lipomyelomeningocele is due to incomplete posterior neuropore closure with associated abnormal separation of neuroectoderm from cutaneous ectoderm. It is characterised by a fatty mass contiguous with the subcutaneous fat and a splayed spinal cord. Patients present with an eccentric skin-covered back mass, often with cutaneous stigmata like hairy patches or dermoid sinus. The patients are typically neurologically normal at birth, but may develop neurological dysfunction if not surgically treated. Imaging features resemble that of myelomeningocele, except that the lesion is eccentrically located, the lipomatous element is present and associated anomalies are less common.⁸ The clinical and radiological features of lipomyelomeningocele are absent in this case.

References

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