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Enteromyeloschisis. A rare Sacro - enteric Mass in the Newborn .

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Introduction

A lumbosacral appendage in a newborn is a source of extreme anxiety for parents. While sacral masses are the most common solid tumours of the newborn (1 in 20-45,000 live births), true caudal appendages (human tails) are rare^{1,2}. Sacral masses are frequently teratomas which are composed of multiple tissues alien to their site of origin, derived from more than 1 germ layer. Rarer pathologies like neuroenteric cysts or true caudal appendages (human tail) have been sparsely reported in literature^{3,4}. We present an extremely rare case of a sacral mass of complete gastrointestinal origin and discuss possible aetiology and differentials. We have coined the term "**Enteromyeloschisis**" to describe this lesion as our search of the English literature did not show a report of any similar case where the tail was made up entirely of bowel.

Keywords: Human tail, **enteromyeloschisis**, lumbosacral mass

Case:

A 3 - day old term female neonate delivered to a 32- year old primiparous mother and presented with a fleshy appendage just on the gluteal cheek of the left buttock. She had been delivered via emergency Caesarean section for a breech presentation. There was perinatal foetal distress , associated with meconium soiled brownish amniotic fluid . APGAR score at birth was 8¹ and 10⁵. The back mass was not pre-natally identified despite 4 ultrasound scans. Birth weight was 2.8kg. There was no foot deformity, no history of jaundice, constipation, seizures or vomiting. On examination, she was afebrile, acyanosed, well hydrated with a flat and normotensive anterior fontanelle and normal muscle tone globally. There was no

respiratory distress, no refusal of feeds or constipation. No features suggestive of a cardiac anomaly was observed. Occipitofrontal circumference was 33cm and no dysmorphic facie was observed.

Musculoskeletal examination revealed active movement of all limbs. There was a right-ward displaced anal dimple and a patent anus. There was a cold, fluctuant firm, skin covered, dusky coloured fleshy swelling over the sacral region just to the left of the midline with ill-defined finger-like projections/ lobulated margins at its lower border(Fig.1a,b). The ventral aspect of the mass, on elevation, had a mucosal plate bearing 2 openings, having the appearance of a vulva and discharging scanty amounts of serous fluid(Fig.2a). The plate was wet and only visible on elevation of the mass. At the supero-lateral portion of the mass arose a bifurcated twig-like fleshy appendage, approximately 6cm from its base. The appendage was pinkish, raw, tube-like, erythematous and cobbled in consistency. There was tenderness on contact with the bifurcated appendage but no contact bleeding.

Digital rectal examination revealed a good sphincteric tone with no palpable rectal mass.

An MRI done on the 2nd day of life before presenting at our facility reported normal spinal cord elements up to the coccyx but with an anterior meningocele from L2 to L5. Also noted was a gas filled, pelvic mass continuous with the spinal canal from the lower sacrum, surrounded by a CSF fluid track, abutting on, but not connected to the rectum with a definite cleft in the musculature (Quadratus Lumborum) allowing exit of the pelvic mass seen outside as the appendage(Fig 3). A urologic evaluation with an abdominal ultrasound reported no abnormality.

She was fed as tolerated, commenced on Vit. K and empirical antibiotics. She had regular bowel movements through the anus. The bifurcated swelling was washed with saline and covered with antibiotic impregnated lubricated gauze dressings. She was prepared for surgery to prevent desiccation and necrosis of the appendage which was anticipated to be bowel because of its length and consistency.

Intra-operative findings were of a weeping T shaped sacral appendage extending into the sacral spinal canal at S5. At its base was a wide based fibrofatty tumour extending to the lumbosacral area.. We made a linear midline skin incision in the normal skin and extended into an elliptical incision around the base of the fibro-fatty mass. Deepening the incision to the subcutaneous layer, we excised the appendage by blunt dissection and with

electrocautery at its origin taking care to dissect off the surrounding skin, spinous process and lumbodorsal fascia. There was a deficient posterior element of the lower sacral vertebrae (S4) but the dura was intact(Fig.2b).The resultant spinal defect was traced to its dural attachment. The cord was also intact and there was no evidence of an anterior meningocele as assessed by the neurosurgeon, contrary to the MRI report. Both masses were amputated with no spillage of fluid or faeces from the cavity. The appendage was blind ending and not in continuity with the rectum. The anal sphincter was stimulated on table with low voltage diathermy and good sphincteric excursion was appreciated. No neurological deficit was demonstrable in the limbs immediate post-op and on follow up for the past 5 months.

She had post operative wound dehiscence on post-op day 5 as there was no full complement of nursing staff available because of the lockdown associated with the COVID -19 pandemic. Dressings change were irregular and compliance with nursing in prone position was poor. She was discharged after 2 weeks and has remained well and active for the past 5 months with no obvious neurological sequelae.

Pathologic examination was reported grossly as a specimen consisting of a single piece of an irregularly shaped grayish white tissue partially covered by skin, weighing 50g and measuring 9x5x3cm. There was an exophytic T-shaped mass attached to the surface that measured 9x3x2cm. The structure appeared tubular, soft to firm in consistency with surface erosion. The skin tissue to which the structure was attached, measured 6x4cm. Cut sections through the structure showed tubular wall with grayish white surfaces and probe patent central lumen. Histologic section showed colonic tissue comprising of mucosa, submucosa, muscularis propria and serosa. The lumen contained amorphous eosinophilic material with scanty inflammatory cells infiltrate. Epithelium and mucosal glands were unremarkable. However there was marked hyperplasia of the muscularis mucosa, and the serosal surface was inflamed with marked oedema, mixed inflammatory cell infiltration, numerous congested vessels and fibrin deposition. The attached skin tissue appeared unremarkable consisting of epidermis, dermis, subcutaneous fatty tissue and skeletal muscle(Fig.4a&b).

Discussion

During the third week of intrauterine development, gastrulation occurs leading to the evolution of the 3 germ layers; endoderm, mesoderm and ectoderm⁵. During this process, there is a complex interaction of primitive cells to form the embryonic precursors of various organs and systems in the body. Tissues derived from the ectoderm include the epidermis ,

brain and nervous system while the endoderm forms the inner lining of the gut among others. The primitive gut tube initially forms a hollowed-out cylinder of endoderm encased in a sheet of mesoderm⁵. After significant elongation, the endoderm then folds cranio-caudally at the ventral portion, bringing both ends together to form a closed tube near the yolk sac⁶. From the migration of cells between the epiblast and hypoblast layers early in the 3rd week, to the formation of the notochordal plate, buccopharyngeal membrane and the cloaca, the complexity and precision of these processes forbids interruption or mishaps. The origin of this spinal anomaly that involves the gut can thus be traced to these early weeks of embryogenesis, probably in the 3rd week to 5th week. The differential diagnoses that were considered pre-operatively in this case included Teratoma, Spinal dysraphism, and Enteromyeloschisis.

We opine that the embryopathogenesis of enteromyeloschisis may be reminiscent/ similar to that seen in neuroenteric cysts, another rare abnormality of the spinal axis composed of heterotopic endodermal tissue⁷. It has been postulated that persistence of the normally transient neurenteric canal impedes appropriate separation of the endoderm and notochord. This abnormal, atypical merger results in congenital anomalies of the spine characterized by presence of mucus secreting epithelium. Neurenteric cysts are allied with bony abnormalities of the spine in approximately 50% of cases and are associated with a variety of conditions including spinal dysraphism, scoliosis, spina bifida among others.⁸

A teratoma was also considered as a differential pre-operatively. Teratomas are tumours originating from totipotential cells and therefore contains tissues from more than one of the three germ cell layers. As a term first applied by Virchow in 1869 to sacrococcygeal masses, the common theories of development of a teratoma include; anomalies of germ cell development, misplaced blastomere, persistence of the primitive streak, incomplete twinning or abnormalities of gastrulation^{9,10}. Totipotent primordial germ cells arise in the 4th and 5th week of gestation from among the endodermal cells of the yolk sac and migrate to the gonadal ridges, and teratomas can develop anywhere in the midline if the germ cells fail to migrate to appropriate target destinations. Teratoma in the human tail has also been reported^{2,11}. In a review series of 48 skin-covered lumbosacral masses, 25% had lumbosacral and sacrococcygeal teratomas¹².

Spinal dysraphism describes a group of congenital anomalies /malformation in the dorsum of the embryo, related to failure of fusion of distal elements. Broadly classified into *occulta* and *aperta*, postnatally, the standard diagnostic modality is radiological (CT/MRI). Occult Spinal

dysraphism often lends itself to suspicion with cutaneous stigmata which include hypertrichosis, lipoma, hyperpigmentation, dermoid cyst or sinus, haemangiomas, large sacral dimple, pseudotails, acrochordon and tails (human or faun)etc^{13,14}. The huge hyperpigmented skin lesion at the base of the appendage and displaced gluteal cleft in our patient was a pointer to an occult spinal dysraphism. Reminiscent of an acrochordon, it was however not localized in a skin fold or in any previously documented classical location (axilla or groin).

The fleshy mass was also not a spinal lipoma, which is associated with about 35% of occult spinal dysraphism. It was made up entirely of epidermal cells of normal skin and there were no cystic spaces within it. The lesion on our patient appeared grossly like a tail. True human tails are described as vestigial remnants of the intraembryonic tail^{15,16}. They are known to contain vascular endothelium or blood vessels, nerves, fatty tissue and connective tissue bundles but are devoid of spinal cord and cartilage. They commonly occur in the lumbosacral area. The histology of the mass in our patient demonstrated only intestinal tissue, with colonic epithelium specifically.

Our patient had no evidence of fused or hemivertebrae but there was a defect in the sacral spine at S4 suggestive of a partial agenesis. The dura was intact and there were no neural elements damaged intraoperatively..

The preoperative assessment of our patient consisted of a detailed pregnancy history, thorough physical examination, magnetic resonance imaging, contrast enema and blood chemistry. All patients presenting with lumbosacral masses must be thoroughly examined, evaluated and investigated. Magnetic resonance imaging is the standard radiodiagnostic investigation in these cases. There was no demonstrable connection of the "tail" with the intraperitoneal viscus during a double contrast enema of the bowel.

Conclusion: We present a rare case of a lumbosacral mass appearing like a human tail, surrounded by exuberant skin and consisting exclusively of gastrointestinal tissue originating from a spinal defect. This is an unusual association of a sacral mass of well-formed intestinal tissue with mild spinal dysraphism that we have coined an **enteromyeloschisis**.

Patient consent: Consent to publish the case report was not obtained though the parents gave verbal consent. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship: All authors attest that they meet the current ICMJE criteria for Authorship.

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(Fig.1a) Lumbosacral mass with appendage



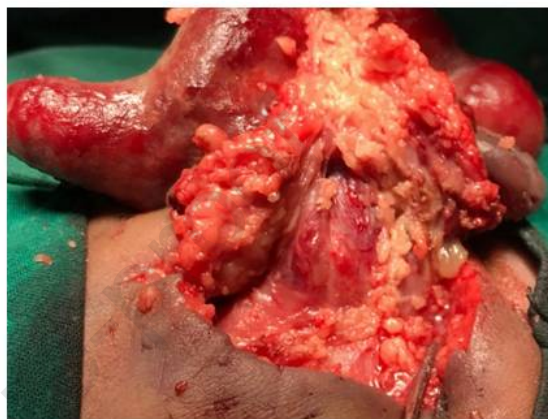
(Fig.1b) Appendage arising from the base of skin covered mass



Fig.2a Vulva/placode like plate at base of tumour



Fig.2b Tumour bed



- (Fig. 3)MRI scan showing appendage exiting the sacral and muscle plane

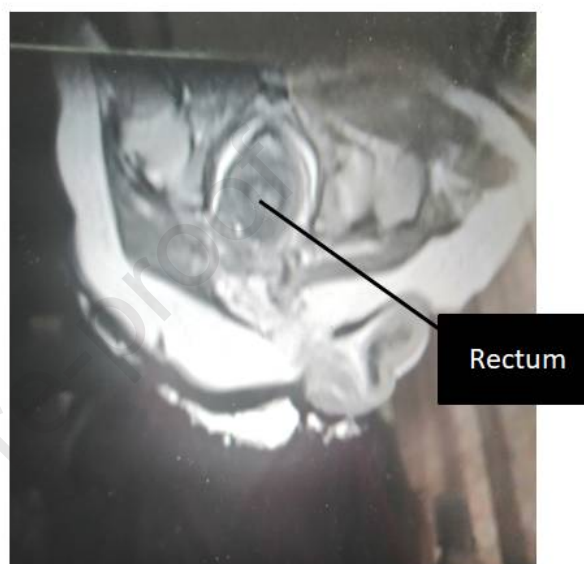
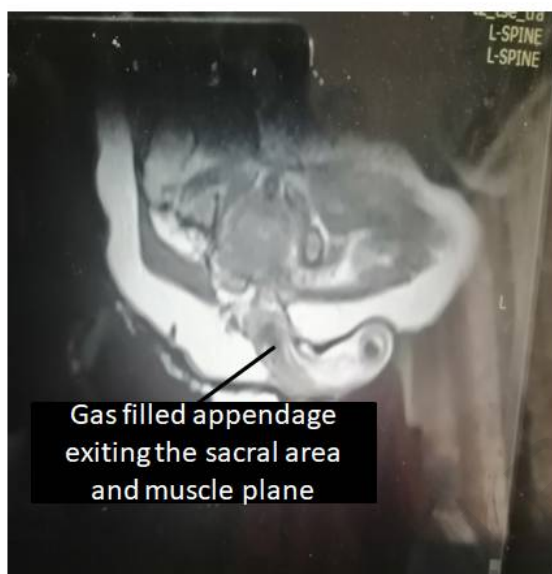
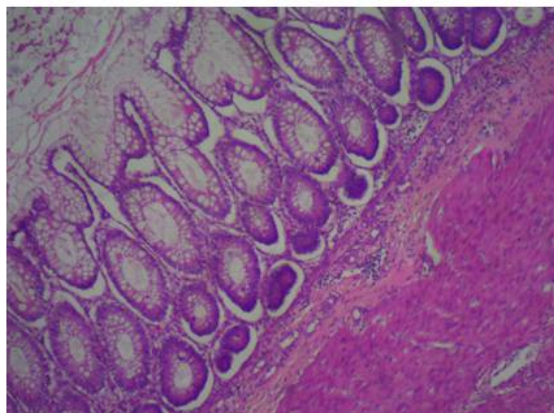


Fig.4a Photomicrograph showing wall of the colon (H&Ex100)



Inflamed serosal layer(H&Ex100)

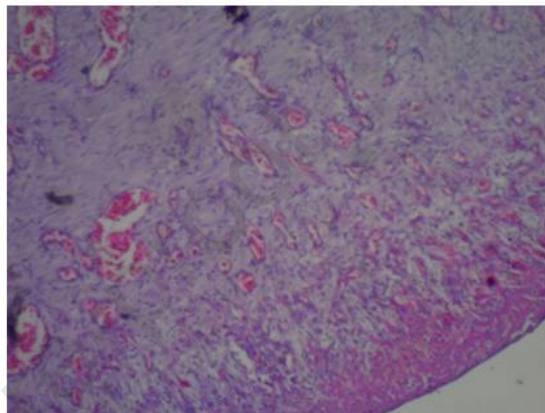
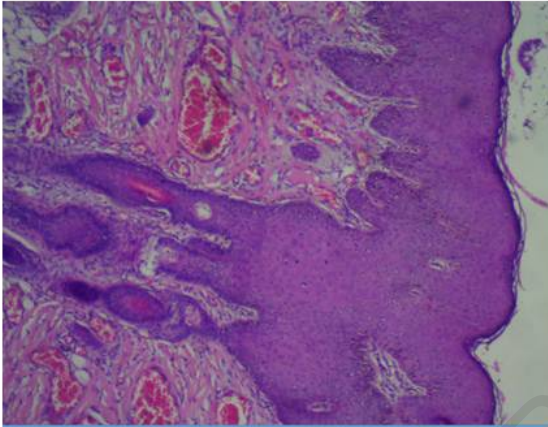
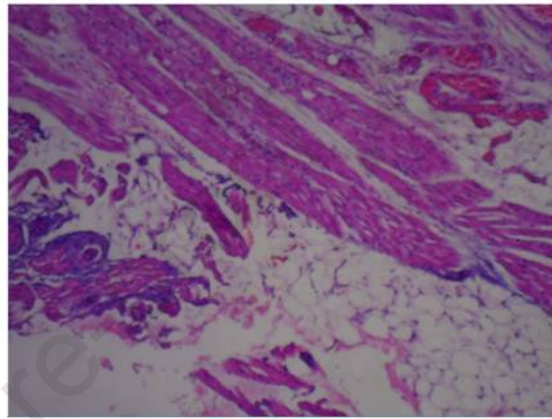


Fig.4b. Attached skin (H&E x100)



Subcutaneous tissue



Declaration of interests

☐ The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

☐ The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: