Case Report

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Parasitic twin within spina bifida

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ABSTRACT

Spinal dysraphisms in association with accessory limbs are extremely rare congenital anomalies. The potential mechanisms of development and classification of these anomalies remain unclear. We report a unique case of a baby boy with an accessory foot and external genitalia in association with spina bifida.

Key words: Heteropagus, myelomeningocoele, parasitic twin, rachipagus, spina bifida

INTRODUCTION

Spinal dysraphisms in association with accessory limbs are extremely rare congenital anomalies. The potential mechanisms of development and classification of these anomalies remain unclear. We report a unique case of a baby boy with an accessory foot and external genitalia in association with spina bifida.

CASE REPORT

A 3-day-old full-term baby boy was referred with an abnormal swelling on the lower back which had a well-developed foot and accessory male genitalia. The baby had been delivered by emergency caesarean section due to prolonged labour and had a birth weight of 3500 g. The mother noted that there was movement of the lower limbs, he was continent and breastfed normally. The mother's antenatal records showed no history of smoking, alcohol, or any regular medication. There was no family history of twinning or congenital anomalies and the marriage was not consanguineous. The mother had suffered from weight loss in this pregnancy, but no retroviral treatment was indicated. She had received a dose of anti-malarial prophylaxis and two doses of tetanus toxoid.

On examination, he was an otherwise healthy baby with a skin-covered swelling in the lumbosacral region, measuring 8 cm by 12 cm, with solid and cystic areas, and containing a well-developed foot and male genitalia [Figures 1 (a) and (b)]. In the absence of a computed tomography (CT) scan, ultrasonography was performed which revealed a mixed solid and cystic lesion in association with a myelomeningocele. There were no associated lipomatous masses. There was no hydrocephalus, and no evidence of cardiac or renal anomalies. A plain radiograph showed a normal lumbar spine [Figure 2]. The patient was scheduled for urgent surgery, when the entire mass was surgically excised, and the myelomeningocele was closed, with an uneventful postoperative outcome [Figure 3].

DISCUSSION

There have been a few reports in the literature of dorsal midline masses containing a variety of well-differentiated germ layer derivatives in association with spinal dysraphism.[1] These have been variously described and classified as spinal hamartomas[2] or mature teratomas[3,4] due to the high level of differentiation present. Others describe them as rudimentary parasitic twins (rachipagus or heteropagus), occupying part of the spectrum of conjoint twinning, ranging from the presence of an internal teratoma, to an externally attached parasitic twin.[5] Yet others have described them as a "disorganization like syndrome".[6]

There has been considerable debate regarding the pathogenesis of these lesions, in particular, those associated with neural tube defects.[7] Krishna and Lal,[1] building on the earlier hypotheses of Gardner and Egar,[1] have proposed a secondary neural tube defect that occurs after primary neural tube closure, caused

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Figure 1: (a and b) Foot and male genitalia on myelomeningocoele

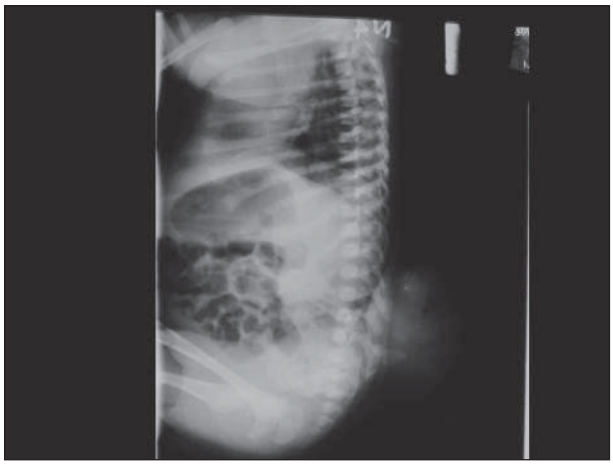


Figure 2: A plain radiograph showed a normal lumbar spine



Figure 3: Post-surgical repair

by over-distension of the already closed neural tube with fluid, and its subsequent rupture, with resulting infiltration of the mesoderm with proteinaceous neural fluid. This appears to lead to a displacement and de-differentiation of Schwann cells to form a mesodermal blastema beneath the intact ectoderm. This gives rise to the development of components of mesenchymal tissue which form accessory limbs in association with midline masses. This "secondary injury" theory may explain why the formation of accessory limbs is more common in association with lipomatous masses, such as lipomyelomengoceles; and less so with spina bifida cystica, which may be a primary neural tube defect.

Their appearance in the setting of developmental spinal cord anomalies suggests a disembryogenetic rather than a neoplastic process. Koen *et al.*[3] postulated that abnormal inductive interactions involving the caudal cell mass occur in intradural spinal teratomas in association with cord malformations, myelomeningoceles and lipomyelomeningoceles. Disruption of differentiation of the pluripotent caudal cell mass itself may be implicated in the development of these spinal cord malformations.

It is hypothesized that conjoined twinning and true duplication of the spinal cord occur at earlier stages of development (at gastrulation or the primitive streak stages) as a result of abnormal upregulation of organizer genes, such as Wnt-8c.[8] The asymmetry seen between the autosite and parasite in heteropagus twinning may be secondary to a vascular insult in the parasite, leading to selective ischaemia of tissues within the parasite, and causing reliance on collateral supply from the autosite. However, vascular insults in other settings such as in placental insufficiency usually selectively spare brain tissue, yet most heteropagus twins are anencephalic.[9] Logrono *et al.*,[10] using fluorescent *in situ* hybridization techniques, found some potential allelic differences between heteropagus twins, and thus proposed that

they may have originated as dizygotic twins, with subsequent fusion. This theory remains controversial, as there are no other reports of digyzosity in heteropagus twins, and the use of fixing and staining techniques could potentially be contributory towards the perceived allelic differences.

The prognosis of these patients remains generally good, more so in comparison to symmetrical conjoined twins, as there are usually fewer shared visceral and vascular structures.[9] Poor outcomes are usually related to comorbidities, in particular developmental anomalies and sepsis. Antenatal diagnosis plays an important role in identifying associated congenital anomalies, which significantly influence prognosis; and in planning obstetric and surgical management. In the absence of antenatal screening, most patients present in infancy, but a few patients may present in pre-adolescence and late teenage years.

Management of parasitic twinning usually involves a single surgical procedure soon after birth to excise the mass in its entirety. Figures from available data on outcomes report mortality in the autosite in 31% of cases (15 out of 49).[9] Cardiorespiratory failure associated with congenital cardiac defects was the most commonly cited cause of death; sepsis was also a major cause of mortality in the context of bowel communications between the autosite and parasite. Morbidity included teratoma recurrence at the site of excision, formation of incisional herniae and iatrogenic injury associated with surgical procedures. There is no specific data with regard to outcomes in children with parasitic twinning in association with myelomeningocele. However, extrapolating from the data above, it is likely that most important determinants of prognosis in these children would be the neurological and urological consequences of the underlying neural tube defect(s).

We report this unusual case of parasitic twin arising with a spina bifida deformity, with a positive outcome in the immediate post-operative period. In this child, we expect the long-term outcome to be favourable, given the initial presentation with limb mobility and continence in association with myelomeningocele.

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