A Very Unusual Case of a Dorsal Heteropagus Twin

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Introduction

- Conjoined twinning is a rare occurrence
- Estimated prevalence = 1:40,000 to 2,000,000
- Southeast Asia and Africa = 1:14,000 to 25,000
- Female : Male ratio = 3:1
Nomenclature

Diplopagus
  • complete, equal and symmetrical

Heteropagus
  • incomplete, unequal and asymmetrical
  • the parasite is smaller and dependent on the host (autosite)
  • parasite is either attached to any portion of the body or even within the host (fetus-in-fetu)
Classification

- according to the most prominent site of union

Thoracopagus
- most common; anterior thoracic conjunction

Rachipagus
- only 2%
- most common site is at the midportion of the posterior vertebral column above the sacrum
Conjoined Twinning

- thoracopagus
- rachipagus
- lumbar rachipagus

DIPLOPAGUS
HETEROPAGUS


Case Report

- 3 year-old male
- born with a dorsal mass
- otherwise asymptomatic

Figure 1. Our patient.
Dorsal Mass

Consists of:

• immobile lower limb in permanent flexion due to soft tissue fusion
• supernumerary toes
• buttocks
• anus-like orifice
• phallic structure above the foot

Figure 2. Our patient with the parasitic rachipagus in lateral (A) and posterior (B) views.
Magnetic Resonance Imaging

AUTOSITE
• syrinx at T5 to T8
• spinal cord tethering below T8

PARASITE
• large predominantly fatty mass from T5 to L5
• no musculature along the bones
• blind-ending external orifice in the parasite
• phallic structure shows no urethra or adjacent testes
• indiscernible soft tissue and cystic components
Figure 3. T1W (A) and T2W (B) sagittal MR images showing the syrinx and cord tethering.
Computed Tomography

AUTOSITE

• spina bifida at T6 down to the sacral bones
• butterfly vertebrae at T11, L1 and hemivertebra at T12 causing levoscoliosis
• thoracic and abdominal cavities are grossly unremarkable

PARASITE

• malformed vertebrae, pelvic bones, femur, tibia, fibula and supernumerary digits are noted
Figure 4. Radiograph, coronal and 3D reconstructions of the bones.
CT Angiography

- Blood supply is from a spinal branch of a right intercostal artery from the descending thoracic aorta at the level of T10 vertebral body
- No discrete draining veins
Figure 5. Axial post-contrast CT images and 3D reconstruction of the blood supply.
During surgery
Histopathology

- Mature elements from skin, bone, cartilage, muscle, fat, brain, respiratory and cervical epithelium were obtained.
- The phallic structure contained several cysts of epidermal origin all lined by skin epithelium (see below).
Discussion

- Most heteropagus twins are either joined ventrally above the umbilicus (epigastric heteropagus) or dorsally in the vertebral column (rachipagus)
- 28 rachipagus (dorsal heteropagus) twins have been reported
- Only 2 are diplopagus, the rest are heteropagus
- 18 females, 5 males, 5 not reported
- 24 (86%) has thoracic and/or lumbar connection
- 9 had meningocele
Discussion

- accurate imaging to define anatomic fusion, vascular and other anomalies and for surgical planning
- CT and MR of the spine are needed for rachipagus type
- assessment of the cardiovascular system is recommended in all types
- no overall mortality or morbidity reported
- outcome better without shared vital organs
- prognosis depends on type and extent of union
Conclusion

Hence, this is a very unusual case of an otherwise asymptomatic boy with a parasitic rachipagus where CT and MRI played a crucial role in the diagnosis, assessment of other anomalies, and for surgical planning and prognostication.
References