Quiz

What is your diagnosis?

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Mrs X, a 26-year-old Primigravida, presented to us for a morphology scan at 20 weeks of gestation. She had been married for 7 months and had conceived following one cycle of ovulation induction with clomiphene citrate. She was diagnosed to have gestational diabetes earlier in pregnancy and was started on diabetic diet following which her sugars were controlled. She had a ultrasound scan (USG) at 6 weeks which showed single live intrauterine pregnancy corresponding to the gestational age. She did not have a history of intake of any teratogenic drugs.

Morphology scan was suggestive of a monochorionic monoamniotic twin gestation. One of the twins was well formed and had normal anatomy and biometry which corresponded to 20 weeks. Attached to upper abdomen and thorax of the normal twin, was a significantly underdeveloped “co–twin” which had only trunk, both lower limbs and rudimentary upper limbs. (Fig1.A)

The scan findings were discussed in details with parents. Risks of surgical separation of conjoined twins; chances of survival of normal twin and the need for Lower segment caesarean section as a mode of delivery in case the pregnancy is allowed to continue, were discussed. The parents opted for termination of pregnancy.

Autopsy findings revealed partial conjoined twins with male autosite weighing 340 gms. The weight of the parasite could not be determined separately. The parasite co-twin was attached to the autosite at the level of epigastrium

The autosite showed patent anus and no significant gross abnormality. The parasite had malformed limb buds, intestinal atresia and poorly developed spine. (Fig 2A,2B) Microscopic cut sections of umbilical vessels showed four arteries and single umbilical vein.

ANSWER

Epigastric heteropagus is a very rare type of conjoined twins. They are also known as asymmetrical or parasitic conjoined twins and it’s a rare complication of Monozygotic twin[1] Their prevalence is 1 in 1-2 million births[2]. The well-formed twin is known as an autosite whereas its under developed counterpart, as parasite, since it is dependant on the former for its growth. The term heteropagus was coined for the first time by Potter and Craig[3]

Asymmetrical conjoined twins are 20 times less common than symmetrical ones [4]. Heteropagus twins are predominantly (78%) males whereas symmetrical conjoined twins are mostly (70%) females. In our case also, both the abortuses were males suggesting monozygotic twinning [5] Symmetrical conjoined twin share bowel and other organs whereas asymmetrical do not share organs [6].

The pathophysiology of the heteropagus twinning has been explained by three theories. The “fission” theory suggests incomplete separation of the embryo [7] while the “fusion” theory proposes coalition of 2 originally distinct parts [7]. The third theory postulates that it occurs due to vascular compromise in utero, leading to death and partial resorption of one of the twins.[7]

Most of the cases described in literature have been diagnosed postnatally [8]. Few, however, were diagnosed prenatally like ours.[9]

The significance of the absence or presence of sharing of organs between the parasite and the autosite is that the surgical separation is less complicated compared with the symmetrical conjoined twins. Since management of such cases is solely based upon case reports, there is still no consensus about mainstay of therapy. When diagnosed early in pregnancy, termination may be a viable option especially in the developing world where surgery required for twin separation may not be widely accessible.

Surgical separation and closure of the incision may be attempted once babies are born. Wound breakdown is a dreaded complication following surgical separation.[10] Long term complications include hernia and teratoma at
the incision site [11] Long term follow up of these babies following separation are very limited. One baby was who followed up to 52 months following surgical separation had normal growth and development [12]

References
**Fig. 1A.** 2D Ultrasound image showing transverse axial view of the upper abdomen of auto site with the stomach bubble. The arrow shows the origin of the heteropagus with trunk, two lower limbs and malformed upper limbs, from the epigastric region of the autosite.

**Fig. 1B.** 3D image of the Epigastric heteropus twin, parasitic twin arising from epigastric region of the auto site.
Fig 2A Gross specimen showing normal appearing male twin arising from the epigastric region of which is the parasitic co-twin. Fig 2B Soft tissue pedicle connecting the parasite to the autosite and its relation to cord insertion on the autosite. Parasitic co-twin was also male with no skeletal muscle in the lowerlimbs.