Congenital Microgastria

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Congenital microgastria is a rare anomaly of the stomach that results from impairment of normal foregut development. The dimunitive stomach lies in the midsagittal plane and does not develop a fundus, corpus, or antrum. It is associated with a spectrum of other anomalies particularly of the intestines and musculo-skeletal system. We document an infant with congenital microgastria with a constellation of associated anomalies.

Case Report

A 10-day-old male child born at term to a primigravida mother of an uncomplicated pregnancy was referred to us for management of lumbar menigomyelocele. Examination revealed an unusual spinal dysraphism with the laminae of affected vertebrae forming a prominent spur by the side of a small meningomyelocele *(Fig. 1)*. He also had dextrocardia which was confirmed by chest roentogenogram. CT Scan and myelogram of lumbar spine revealed presence of diastematomyelia and tethering of cord along with a small meningomyelocele.

To rule out the possibility of situs inversus, an upper gastrointestinal contrast study was performed which revealed a small saccular stomach lying in the midsagittal plane having no discernible fundus, corpus or antrum *(Fig. 2)*. The esophagus was dilated and there was significant gastro-esophageal reflux. Ultrasonography of abdomen confirmed the midsagittal position of stomach; there was no evidence of situs inversus and the spleen was enlarged.

Excision of spur causing diastematomylia, untethering of cord, and excision-repair of meningomyelocele using free fascia lata graft was done. The post-operative period was uneventful.

The baby had frequent regurgitation of feeds, which improved with conservative measures. The child is on frequent small feeds and is gaining weight.

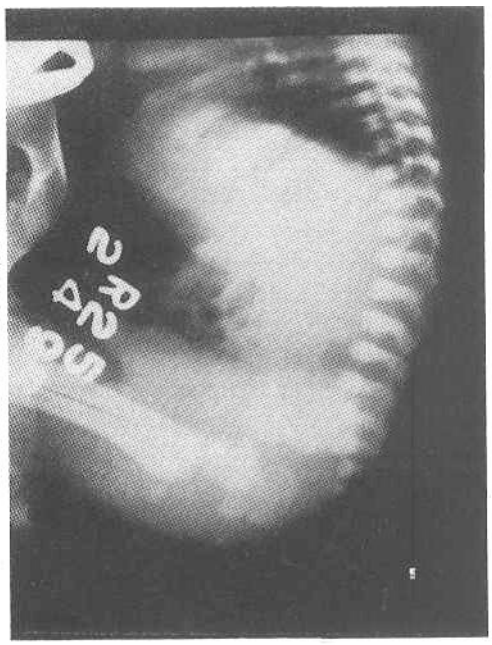

Fig. 1. Lateral view of spine showing the abnormal bony spur formed by dysplastic vertebral posterior laminae overlying the meningomyelocele.

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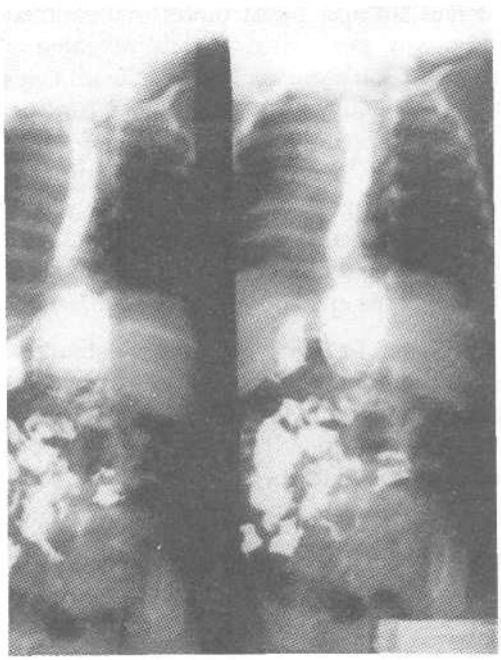

Fig. 2. Upper gastrointestinal contrast study shows a small tubular stomach and dilated esophagus. Intestinal malrotation is present with lack of normal duodenal sweep.

Discussion

Congenital microgastria is an extremely rare anomaly. There are less than 30 previously reported cases in more than 100 years, since the entity was first described by Dide in 1894(1,2). It is characterized by a small, saccular or tubular stomach with minimal reservoir capacity, megaesophagus and incompetence of cardia(2).

Several other foregut and hindgut anomalies have been reported in association with microgastria including esophageal atresia, pyloric atresia, duodenal atresia, malrotation. Hirschsprung's disease and imperforate anus(3-5). Congenital cardiac, faciomaxillary, central nervous system and musculoskeletal anomalies have also been reported(4,5). Although lumbosacral vertebral anomalies have been reported in the past(3), spinal dysraphism in the form of spina bifida cystica has never been reported before. The non-random association of involvement of various organ systems suggests that the anomaly is caused by an aberrant mesodermal involvement in the early embryonic life.

The clinical manifestations vary from patient to patient depending on the stage at which the development of the stomach is arrested. The usual presentation is that of gastroesophageal reflux, postprandial vomiting, malnutrition, and recurrent aspiration pneumonia(2).

The treatment of congenital microgastria must be individualized. In patients with lesser degrees of gastric aplasia, as seen in our patient, conservative management with small frequent feeds may suffice. Bypass or feeding jejunostomies or gastric augmentation surgeries such as creation of Hunt-Lawrence pouch have been described in those having more severe dysgenesis(2,3,5).

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