CASE REPORT
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HEPATIC OMPHALOCELE IN AN ADULT

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Abstract. The finding of an untreated omphalocele in adulthood is extremely rare. We report the case of a 29-year-old patient, who presented to us with a congenital defect of the abdominal wall and protrusion of underlying viscera.

Key words: abdominal wall defects, omphalocele, staged abdominal wall reconstruction.

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An omphalocele is a congenital defect that affects the development of the abdominal wall in the umbilical region with a hernial type sac of variable size. It has an incidence of between 1/3000 to 1/10 000 live births (1, 3, 11, 13, 17). It is generally treated surgically at birth, either as an emergency or as a planned procedure, depending on the specific case and on the malformations associated with it. The type of treatment depends on the size of the omphalocele and on the general condition of the patient, from single-intervention techniques to multistage procedures.

CASE REPORT

A 29-year-old woman presented with an umbilical hernia that forced her to wear a surgical truss permanently.

On examination she had a congenital malformation of the abdominal wall with protrusion of underlying viscera, which we considered to be an omphalocele that had been partially treated in childhood. However, the lack of specific clinical documentation and absence of family who would have been able to help with the history did not permit a clear understanding of either the course of the illness or the previous therapeutic interventions.

Examination

Clinical examination of the abdominal region showed an elliptical area along the midline where the abdominal wall was incompletely formed, which extended longitudinally for 18 cm from the xiphoid process to the hypogastrium and transversely for 9 cm between the two pararectal lines, with no umbilical scar. The viscera beneath were palpable and covered by a thin, hypotrophic cutaneous layer. The structure of the thoracic arches was visibly asymmetrical, resulting in a difference of position and shape between the two breasts (Fig. 1*a*).

Computed tomograms of the abdominal region showed hypoplasia and diastasis of the rectus abdominis muscles in the mesogastric zone; the liver was enlarged and dysmorphic, lying immediately beneath the anterior abdominal wall with partial herniation at the superior part of the diastasis. The spleen was displaced in the left iliac fossa and the kidneys, although normal in shape and position, were moderately enlarged.

No interatrial cardiac or vena caval anomalies were found. The intestinal loops were distributed irregularly, with the small intestine located in the right quadrant and the large intestine in the left. After contrast medium had been given a slowing of intestinal peristalsis was shown, as well as opacification of the caecum and of the ascending colon, which looked to be in the midline within the pelvis. Radiographic examination of the thorax showed serious scoliosis with the convexity to the right and marked asymmetry of the two costal arches.

Diagnosis and treatment

An omphalocele with no associated anomalies was diagnosed, that had been partially treated in childhood

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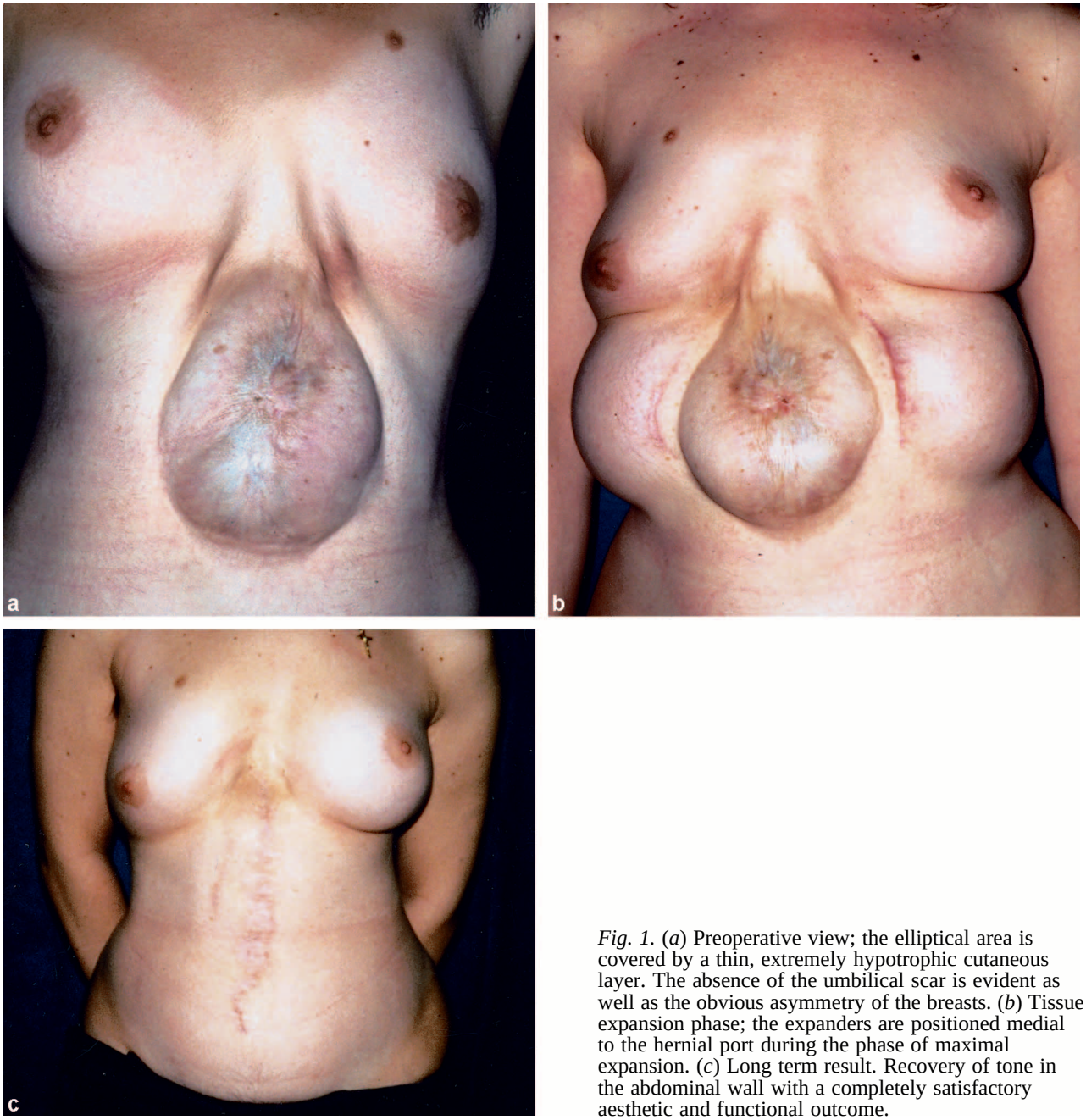

Fig. 1. (*a*) Preoperative view; the elliptical area is covered by a thin, extremely hypotrophic cutaneous layer. The absence of the umbilical scar is evident as well as the obvious asymmetry of the breasts. (*b*) Tissue expansion phase; the expanders are positioned medial to the hernial port during the phase of maximal expansion. (*c*) Long term result. Recovery of tone in the abdominal wall with a completely satisfactory aesthetic and functional outcome.

with cutaneous covering of the hernial sac (Gross Technique) (12). The defect was repaired by closure of the fascia and muscles of the deep layers using a mesh, and the use of tissue expansion to achieve continuity of the skin.

The operation was done in two stages:

During the first stage two rectangular expanders, 16 cm by 9 cm with a volume of 500 ml, were placed at the level of the flanks, lateral to the hernial port. After she had recovered she had regular filling of these expanders as an outpatient until expansion of 15% of their volume capacity had been reached, to obtain sufficient skin for closure of the abdominal defect (Fig. 1*b*).

During the second stage the abdominal wall was reconstructed. The expanders were removed and the dystrophic cutaneous tissue covering the visceral protrusion was excised. The peritoneum was carefully detached from the overlying dermal layers through a midline vertical incision avoiding the xiphoid, so allowing direct exploration of the abdominal viscera and their repositioning within the cavity.

The peritoneum was then closed and the abdominal wall reconstructed in layers. The previously saved

dermal layer was used to form a first layer, and a mesh of polypropylene was positioned over it to cover the entire hernial gap. The two rectus abdominis muscles, which were small and considerably displaced laterally from their points of origin, were identified and isolated. The muscular fascia was sutured medially forming the third layer. The two rectus abdominis muscles were then mobilised and sutured medially to reinforce the reconstruction of the abdominal wall. Continuity of the integument was ensured by bringing the expanded cutaneous tissue together medially without tension.

The patient was discharged after four days recovery and is well after four years of follow up. The result remains satisfactory from anatomical, functional, and aesthetic points of view (Fig. 1*c*).

DISCUSSION

This case is of interest as it is an unusual pathological finding in adulthood and it has therapeutic problems.

The patient was living an apparently normal life but the congenital defect of the abdominal wall with the associated omphalocele was troublesome. As well as causing aesthetic problems, the protrusion of viscera covered by a thin layer of skin was impinging on everyday life and relationships with the opposite sex, as she was forced to wear a permanent surgical truss to avoid gastrointestinal complications. Correction therefore had both functional and aesthetic objectives.

When planning the treatment, research into the congenital abnormalities that may be associated with the omphalocele was of particular importance (including horseshoe kidney and interatrial septal defects). Specific attention was paid to possible involvement of the inferior vena cava, because, as has been reported, these anomalies are frequently associated with, and are determinants of, the development of severe post-operative complications (2, 10).

Large defects of the abdominal wall with herniation of the viscera and inadequate availability of covering soft tissue, are difficult to treat (5, 6, 14). The basic objectives are: adequate protection and covering of the abdominal viscera, repair and prevention of visceral protrusion, and achievement of an acceptable aesthetic result. Numerous methods have been described, depending on the presentation. The repair of the deep layers by plastic reconstruction of fascia and muscles, in association with polypropylene or polytetrafluoroethylene meshes and the closure of the integument by direct approximation of the cutaneous flaps along the midline is possible, but only for small defects (1, 15, 17).

In the more serious cases such as ours, it is necessary to use other techniques (7–9, 12, 15–17). The method we preferred involved reconstruction of the covering layers using tissue expansion and repair of the deep layers with polypropylene mesh and the closure of fascia and muscles.

We think that using well vascularised tissue, that causes no tension on the sutures, such as expanded tissue (in agreement with Paletta et al. (14), Byrd and Hobar (4), and Carlson et al. (5)) reduces complications linked to the use of prosthetic mesh and guarantees long term stability.

In this case the choice was obviously appropriate, allowing both functional and aesthetic objectives to be reached. Follow-up checks up to four years from the first intervention, have shown optimal recovery of the tone of the abdominal wall and a satisfactory aesthetic result that allowed the patient to resume a normal life.

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