CLINICAL CASE

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Sacrococcygeal Teratoma with a Lumbosacral Intradural Extension – Case Report

Potworniak krzyżowo-ogonowy z zajęciem odcinka krzyżowo-lędźwiowego kanału kręgowego

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Abstract

A case of intradural extension of type I sacrococcygeal teratoma in a full-term newborn is reported. The true anatomical extent the tumor was noted intraoperatively. Following sacral laminotomy, an intradural lumbosacral portion of the tumor was dissected and excised along with its external part. Histological examination revealed a mature teratoma. Postoperative course was uneventful. Normal development without neurologic impairment were noted on follow-up studies over 1 year after excision. This is a fourth case of such unique sacrococcygeal pathology documented in the literature to date. Despite its rarity, this unusual anatomical variant of type I sacrococcygeal teratoma should be considered in differential diagnosis of sacrococcygeal tumor (**Adv Clin Exp Med 2006, 15, 1, 193–197**).

Key words: sacrococcygeal tumor, intradural extension, teratoma, newborn.

Streszczenie

Autorzy przedstawili typ I potworniaka krzyżowo-ogonowego u donoszonego noworodka z wnikaniem w obręb kanału kręgowego. Zasięg anatomiczny guza został stwierdzony śródoperacyjnie. Zabieg operacyjny polegał na radykalnym wycięciu zasadniczej zewnętrznej części guza oraz części śródkanałowej. Badanie histologiczne wykazało utkanie potworniaka dojrzałego (*teratoma adultum).* Przebieg pooperacyjny był niepowikłany. Badania kontrolne wykazały prawidłowy rozwój fizyczny dziecka. Dotychczas w piśmiennictwie opisano zaledwie trzy podobne przypadki guza krzyżowo-ogonowego z wnikaniem w obręb kanału kręgowego. Ten rzadko spotykany rodzaj anatomiczny potworniaka powinien być uwzględniony w diagnostyce różnicowej guzów krzyżowo-ogonowych u noworodka (**Adv Clin Exp Med 2006, 15, 1, 193–197**).

Słowa kluczowe: potworniak, guz krzyżowo-ogonowy, noworodek.

Widely accepted classification of sacrococcygeal teratomas (SCTs) is based on their anatomical extent and intrapelvic topography. The most frequent type I of SCT denotes predominantly external tumors with only a minimal presacral portion [1]. Preoperative diagnostic studies in newborns affected by a protruding sacrococcygeal mass are basically aimed in determining its potential intrapelvic extension. Although association of teratoma and spinal dysraphism have been reported in the literature, involvement of vertebral canal by type I SCTs is an unique pathomorphological feature [2, 3]. An extensive literature search has revealed only three such cases reported in newborns [4–6]. Therefore the patient with type I SCT and intradural extension reported herein is the fourth documented case of such unusual anatomical variant.

Case Report

A full-term female infant was delivered vaginally at 40 week of gestation. She was a second child of a 33-year-old mother with no reported med-

ical problems during pregnancy. Family history was unremarkable. The newborn was in stable condition and immediately after delivery a large soft-tissue mass was noted in the sacrococcygeal area. No other anatomical defects were recognized. Neurological examination did not reveal any abnormality. The baby was referred to the Department of Pediatric Surgery on the first day of life.

Physical examination showed a club-shape tumor measuring 10 × 6 × 5 cm (Fig. 1). The mass was covered by a thinned, partially reddish skin. On palpation it presented a mixed solid and cystic consistency. In a supine postion of the newborn, the tumor covered the perianal and pudendal area with its anterior surface. A distance between its posterior border and the anus, located in a normal anatomical position, was 1.5–2 cm. Ultrasound scan confirmed a heterogenous nature of the tumor with solid and cystic components and without pelvic extension. On plain radiograph in lateral view the mass presented as an uniform opacification without calcifications. An antero-posterior film did not show evidence of sacral abnormality (Fig. 2). Presumptive diagnosis was sacrococcygeal teratoma of type I. Preoperative serum α-fetoprotein (AFP) level of 12 629.30 ng/ml was noted. Ultrasound scan of the head and abdomen did not show any associated abnormalites of the internal organs.

Surgery was performed on day 3 of life. From a posterior approach the exterior portion of the tumor was dissected circumferentially. It was separated carefully from the rectum adjoining closely to its antero-lateral right portion. The coccyx was dissected in "one bloc" with the mass. On both sides of the narrow basic portion of the tumor firm ossified structures were palpated. They were found to form a widened sacral hiatus. Presacral space was not involved by the tumor. An attempt to dissect posterior margin of the basic portion of the tumor revealed surprisingly that it penetrated upwards and invaded into the sacral vertebral canal (Fig. 3). Intravertebral and coccygeal exophytic portions formed a widely angulated V-shaped structure with a remarkably narrowing at the site of their junction. Such unexpected findings prompted an upper sacral laminotomy. The intradural portion of the tumor extended up to the L5 level and filled densely the vertebral canal. Its uppermost portion showed a soft solid consistency. After partial mobilisation and division of posterior thin fibrous attachments, neural elements of the filum terminale and sacral roots were identified on the anterior surface of the mass and very delicately dissected. The tumor was removed from the vertebral canal and excised along with the coccyx. Its intraspinal portion measured 7.5 cm in length (Fig. 4). Following careful inspection, the dura was closed in a watertight manner. The fascio-muscular flaps were approximated in the midline over the lumbo-sacral portion of the vertebral canal. Gluteal musculature was reconstructed. The wound was closed in layers with interrupted sutures after placement of a Redon drain. Postoperative course was uneventful. The wound

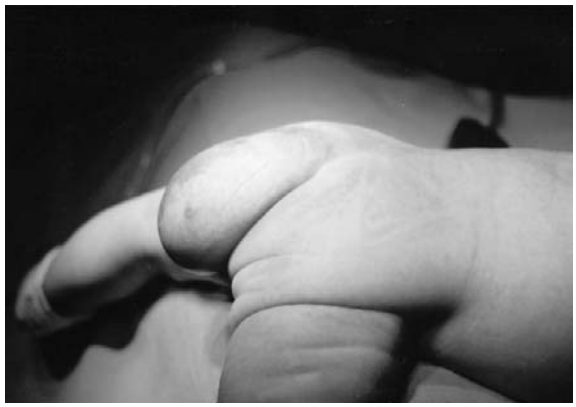

Fig. 1. Preoperative appearance of the sacroccygeal tumor

Ryc. 1. Obraz przedoperacyjny guza krzyżowo-ogonowego

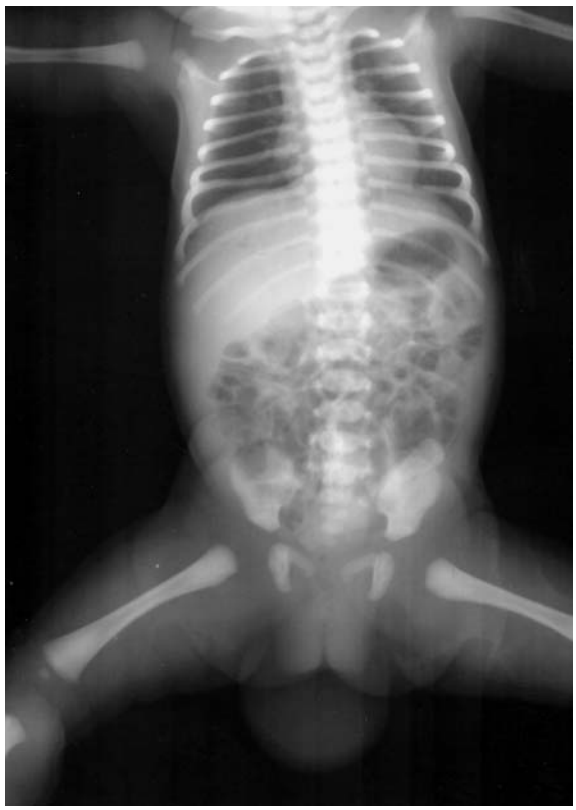

Fig. 2. Preoperative radiograph of the abdomen and pelvis (antero-posterior view). Midline oval opacification without calcification visible

Ryc. 2. Badanie radiologiczne (projekcja AP) jamy brzusznej. Widoczny owalny cień guza ze zwapnieniami

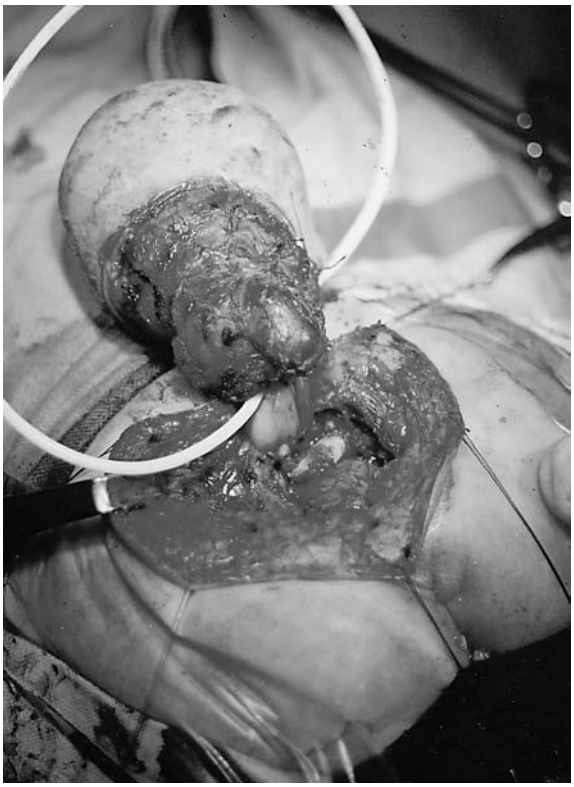

Fig. 3. Intraoperative view of the tumor. The bulky external part was dissected and rotated upwards. A whitish narrow midportion of the mass enters the vertebral canal

Ryc. 3. Obraz śródoperacyjny guza. Część zewnętrzna została wypreparowana i wyniesiona ku górze, część środkowa – zwężona, wnika w obręb kanału kręgowego

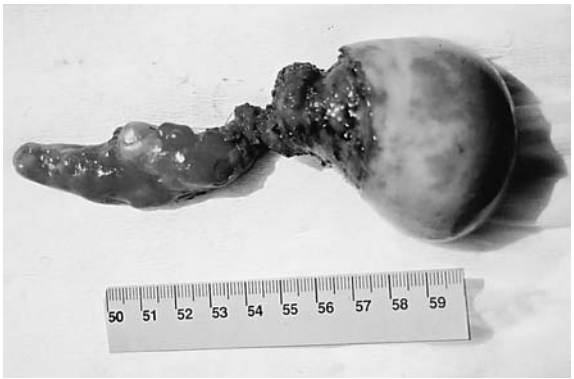

Fig. 4. View of the resected tumor encompassing the external part and its intradural extension

Ryc. 4. Preparat pooperacyjny guza

healed satisfactorily. The patient was discharged from the hospital on 11th postoperative day. Postoperative level of serum AFP was of 3 852.97 ng/ml.

Histological examination of the resected specimen showed a benign mature teratoma with well differentiated components.

On further follow-up spanning over 14 months after the surgical excision the child showed satisfactory physical development. Neurological status was normal with no impairment of lower limb function. Subsequently performed routine biochemical studies did not showed abnormal level of serum α-fetoprotein. There was no evidence of tumor recurrence on ultrasonography.

Discussion

In spite of a great morphological variability of sacrococcygeal teratomas they can be catogorized into 4 anatomical variants depending of an extent of its pelvic component. Most reports from pediatric surgical centers clearly indicate predominance of type I of SCT's among all affected newborns [7–9]. These tumors are easily recognizable immediately after birth and even when characterized by a huge dimensions, they are limited mainly to the sacrococcygeal and gluteal area with no intrapelvic or presacral space involvement. Intraspinal extension of type I SCT is an extremely rare phenomenon. Gross et al reported as first such variant of SCT in 2 newborns but they did not give any anatomical details of the tumor itself [10]. First description of type I SCT with extension into the spinal canal was presented by Teal et al. [6]. Their patient had a sacral mass diagnosed by antenatal ultrasound at 30 weeks of gestation. The infant was born by cesarean section and on day 2 underwent resection of the 12-cm tumor which invaded the spinal canal through the widened sacral hiatus and was attached to the filum terminale. Histological examination revealed immature teratoma of grade 2. Powell et al. reported in details a case of intradural extension of type I mature SCT in a full-term female newborn. An unusual anatomical extent of the 12 × 13 × × 4,5 cm sacrococcygeal tumor was determined intraoperatively. Intraspinal portion of the tumor was attached to the filum terminale and its resection necessitated sacral laminectomy. The postoperative course was complicated by the wound dehiscence with subsequent CSF leakeage [4]. The third case was described by Ribeiro et al. They reported a full-term female infant with a huge sacrococcygeal mass measuring 15 × 10 × 10 cm with intradural and extramedullary extension up to the level of T4. An anatomical extent of the tumor was determined preoperatively by ultrasonography, computed tomography and magnetic resonance. Imaging studies revealed a widening of the thoracolumbar portion of the veterbral canal and a lumbosacral posterior arch defect. Surgical management consisted of a complete resection of the tumor following sacral, lumbar and thoracic

laminotomy. The resected tumor proved to be a mature teratoma with a mixture of nervous and glanular element within its extramedullary portion. On follow-up studies the patiens showed no neurologic deficits but a poor compliant urinary bladder with vesicoureteral reflux was noted at age of 2 years [5]. Reviewing four cases of unusual anatomical variant of SCT with intraspinal extension, including the patient reported above, it should be emphasized that in all of them vertebral anomalies were noted intraoperatively. In three patients sacral bone defect was missed preoperatively, but in these cases preoperative imaging was based on X-rays and ultrasound scan only. Sacrococcygeal tumor invaded the vertebral canal through the posterior sacral or lumbosacral defect [4, 6]. An incidence of lumbosacral spina bifida occulta among babies with an SCT has been reported between 5–15%. Lahdenne et al. performed follow-up study in adults who had previously undergone surgical management of SCT [11]. Among 21 patients with type I SCT, they found radiologic evidence of hypoplastic sacrum in 4, spina bifida occulta in 5, and hypoplasia of L5 vertebral arch in further 5 of them. Even higher incidence of various vertebral defect was noted among patients with other anatomical types of SCT. In view of these observations, an extremely rare occurrence of type I SCT with an intraspinal extension must be surprising and seem to indicate a complex pathogenesis of this congenital anomaly. It may be speculated that hypothesis indicating a random association of dysraphic spinal defect and sacrococcygeal tumor with coincidental and secondary affection of the intravertebral space by an ingrowing external tumor does not seem true in this peculiar pathology. Sutton suggested that the tumor might have originated from the occult dysraphism and secondarily formed the predominant exterior sacrococcygeal mass [5]. Such hypothesis fits well to the case reported by Ribeiro et al. In three cases, the intravertebral portion of the tumor was found to be attached to the filum terminale. In only one patients, mixed solid and cystic mass invaded the intradural and extramedullary space but intraoperatively no firm attachements between the tumor and spinal cord were noted. Such intravertebral anatomy of SCT in all four patients suggest rather a slow ingrowth of the tumor without significant disturbance of spinal cord development. It seems additionally that invasion of the intradural space in patients with sacral extension of the SCT may be regarded as a relatively late event. In all but one patients no neurologic deficits were noted on follow-up studies indicating preservation of intact elements of the spine despite presence of an intradural "space occupying" lesion.

An accurate anatomical extent of the mass and verterbral anomaly were detected unexpectedly during an operation in three babies [4, 6]. In the fourth case of extensive involvement of the spinal canal, presented by Ribeiro et al., was the diagnosis made preoperatively [5]. It should be underlined that despite a very rare incidence of intradural extension of SCT, such possibility should be taken into account by any surgeon responsible for the care of patients with sacrococcygeal tumor. Even in cases of type I SCT, imaging studies, apart from conventional radiography and ultrasound scan, should ideally include MRI study in order to determine a full anatomical extent of the tumor, its topography, and to detect all potentially associated defects. More detailed diagnostic imaging should be without doubt indicated in babies affected by SCT, regardless of anatomic type, in whom preoperative radiograph suggests any vertebral anomaly.

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