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CASE REPORT

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Congenital abdominal dumbbell fashion neuroblastoma with invasion of spinal canal detected by ultrasonography – case report

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Summary

Background: A case of congenital abdominal dumbbell fashion neuroblastoma with invasion of the spinal canal detected by ultrasonography (US) is presented.

Case report: A 3-week-old male neonate was admitted to the hospital with a palpable mass in the left lumbar region. Ultrasound examination was performed on the same day. It disclosed a pathologic mass filling the left side of the retroperitoneal space - displacing laterally and inferiorly the left kidney. The second part of the tumor was located above the Gerot's fascia in the muscles and infiltrated the spinal canal via lumbar intervertebral foramens. Computer tomography scanning confirmed the presence of solid masses in these locations. Urinary excretion of vanillin-mandelic acid (VMA) was within normal range, ferritin level was elevated (447 μg/ml). Bone scintigraphy showed metastases to the left clavicle. There were no changes in bone marrow. Diagnosis of an undifferentiated malignant neuroblastoma was established in histopathological examination.

Conclusions: Spinal ultrasonography is highly recommended in neonates and infants with retroperitoneal tumors.

Key words: abdominal neoplasm • congenital • neuroblastoma • spinal cord neoplasm • ultrasonography

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Background

Neonatal solid tumors are very rare, ranging from 1.56 to 3.65 per 100 000 live births. The most common tumor of this period is neuroblastoma (NB) with estimated incidence of 0.61 per 100 000 live births (28-30 % of all tumors) [1].

The first description of a neuroblastoma was given by Virchow in 1864. These tumors consist of one or more components: undifferentiated neuroblasts, differentiating neuroblasts in various stages of maturation, neurophils, ganglion cells, mature neuritic processes, fibrous tissue [2]. The echostructure of the tumor may be solid (56%) or cystic (44%) [3]. The localization of the tumor are peripheral sympathetic nervous ganglia, mostly placed in the adrenal medulla (93% of neonatal neuroblastoma cases) [3]. However, it can be localized in the cervical, thoracic, abdominal, pelvic, spinal ganglia as well.

In 1927, Cushing and Wolbach first reported a 2-year-old child with a dumbbell tumor of the spine [4], however, intraspinal infiltration in the neonatal period occurs in a marginal number of cases.

The paper reports another case of neonatal abdominal "dumbbell" neuroblastoma with invasion of the spinal canal detected by ultrasonography.

Case report

A male neonate was born vaginally at term, without antenatal problems. Apgar score was 10 at the first and the fifth

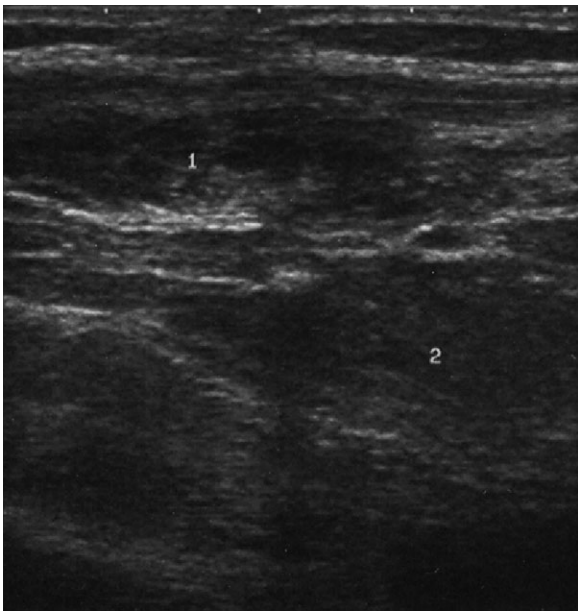

Figure 1. Two pathologic masses of the tumor in the retroperitoneal space.

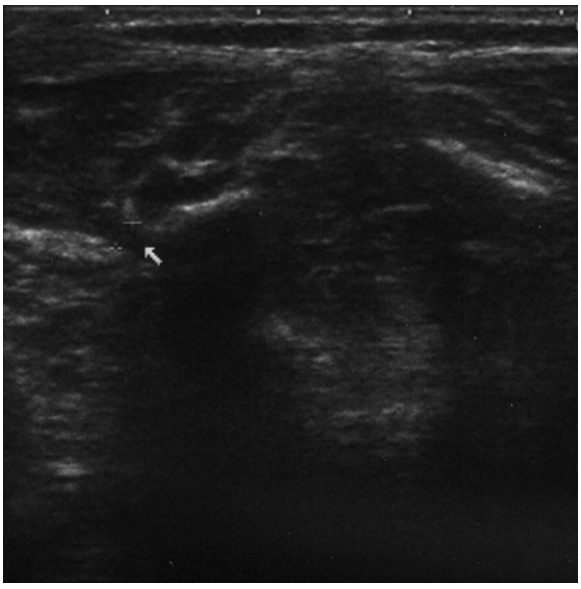

Figure 2. Axial ultrasound scan shows the part of paraspinal tumor extended in a dumbbell fashion into the vertebral foramina (white arrow).

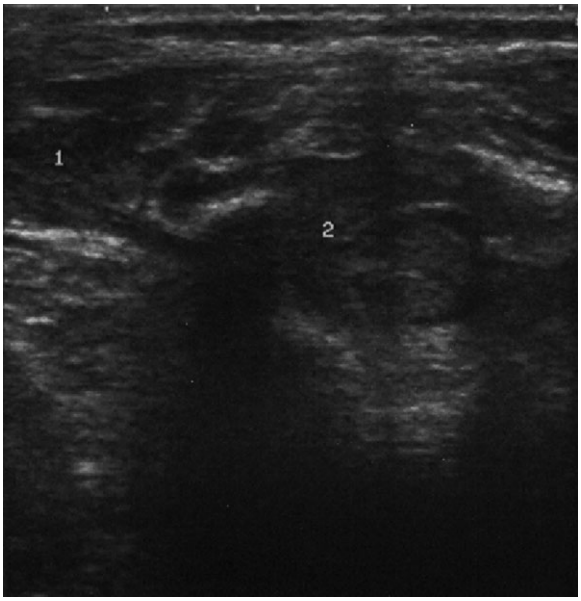

Figure 3. Transverse ultrasound scan shows pathologic mass of tumor located above the Gerot's fascia in muscles (1) and tumor extension to the spinal canal (2).

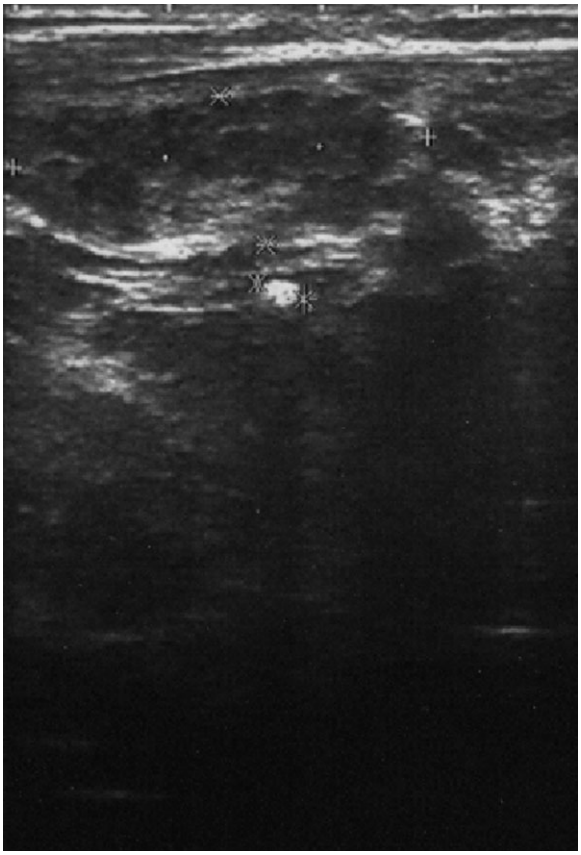

Figure 4. Coronal ultrasound scan shows mass in retroperitoneal space with calcification.

minute. During the gestation, only one ultrasound examination was performed at the 22nd gestation week, with no abnormalities found. There was no risk of infection or adaptation problems at the time following the delivery. After three weeks, the child was admitted to the regional hospital, because of appearance of a palpable mass in the left lumbar region.

During US examination performed in the reference hospital, a solid tumor consisting of two pathologic masses was noticed. The first mass, 44x25 mm in size, was localized in the retroperitoneal space, between the left kidney and the spinal column, shifting the left kidney laterally (Fig. 1). The second pathologic mass, 37x12 mm in size, was located above the Gerot's fascia adjacent to the psoas muscle (Fig. 1). This part of the paraspinal tumor extended in a dumbbell fashion into the vertebral foramina and caused cord compression (Fig. 2, 3).

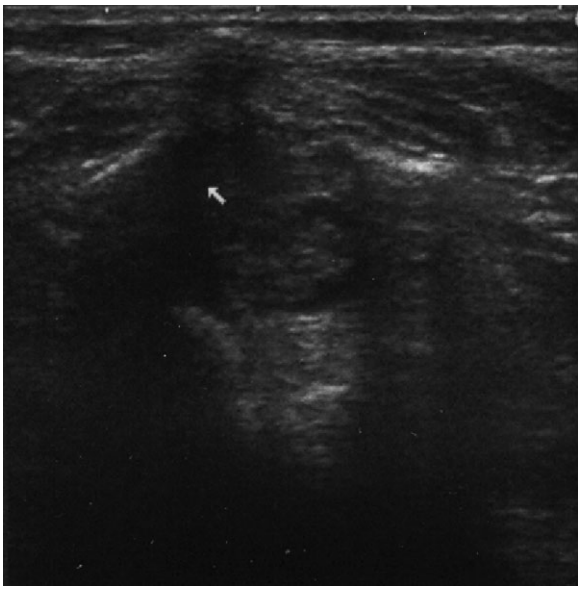

Figure 5. Transverse ultrasound scan shows intraspinal part of the tumor with conus medullaris compression.

All parts of the tumor were solid, normoechoic structures with small hypoechoic foci. There were some calcifications; the largest was 4.8 mm in size (Fig. 4). In the spinal canal, the tumor shifted the spinal cord to the left and invaded its lumbar segment, the conus medullaris and the cauda equina (Fig. 5). Color and power Doppler sonography showed a few small vessels localized around retroperitoneal part of tumor. No Doppler signal was noticed in the spinal canal part of the tumor. The tumor came out at the left side of the adrenal gland. The right adrenal gland, kidneys, liver, spleen, aorta, vena cava inferior were normal and lymph nodes were not enlarged.

Computed tomography confirmed the presence and localization of the solid mass. Urinary excretion of vanillin-mandelic acid (VMA) was within normal range, the level of ferritin was elevated (447 μg/ml; norm 7-282 μg/ml). Bone scintigraphy revealed metastases to the left clavicle. There were no features of bone marrow invasion. Neurological examination revealed shortening and slimming of the left limb, decreased muscle tone of both lower extremities with increased spinal reflexes. Histopathological examination of the tumor established the diagnosis of undifferentiated, malignant neuroblastoma.

Discussion

Congenital abdominal "dumbbell" neuroblastoma is a rarity. However, neuroblastoma is the most common malignant tumor in newborns. Although this form of neuroblastoma was first described in 1927 year by Cushing and Wolbach [5], there are only a few reports in the Medline database up to the present [5, 6, 7].

Radiological assessment is recommended for all newborns and older children with an abdominal mass. An abdominal CT scan or MRI usually follows ultrasound examination [8].

While MRI is the primary modality for the spine imaging in children and adults, ultrasonography is a very useful imaging method in the neonates and young infants [9]. High-frequency linear-array transducers and an excellent acoustic window (the non-ossified posterior elements of the newborns and infants) provide first-rate detail of the spine from the posterior approach [10, 11].

Some authors emphasize the possibility of visualizing a neuroblastoma and tumor vessels even in prenatal examinations [12, 13], and the tumor has been discovered as early as at the 19th week, although the mean age at discovery is the 36th week [13].

In our case, the first ultrasound examination was performed in the 22nd week of gestation and no abnormalities were detected. Unfortunately, US was performed only once during the pregnancy. The second US examination was taken in the 3rd week of the neonate's life. It detected a tumor in the retroperitoneal space and enabled the identification of a local invasion of neuroblastoma to the spinal space with anatomical implications (infiltrations of the lumbar segment of the spinal column, shifting the cauda equina) and clinical ones (neurological deficits).

In neonatal neuroblastoma, the survival rate is greater than 90% [14]. However, in "dumbbell" neuroblastoma fashion, full recovery depends on neurological deficits induced by the duration of spinal cord compression [15]. For that reason, an early diagnosis of congenital "dumbbell" neuroblastoma may have strong impact on quality of future life of the survived children.

Conclusions

Performing spinal ultrasonography in neonates and infants with retroperitoneal tumors is highly recommended.

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