Nonmyxoid mediastinal lipoblastoma in a 2-year-old girl: Case report with US, CT, and MRI findings

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KEYWORDS

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imaging.

Abstract Lipoblastomas are rare, benign tumors of mesenchymal origin that contain adipose tissue. They usually develop in the soft tissues of an extremity in infants, children, and young adolescents. We report the case of a 22-month old girl referred to our staff for swelling in the supraclavicular fossa, which was observed when the child cried. The ultrasonographic examination revealed a mass in the anterosuperior portion of the mediastinum that extended into the right supraclavicular fossa. The lesion was weakly hyperechoic with clear-cut margins and did not appear to invade surrounding tissues. On CT, it appeared inhomogeneously hypodense with denitometric characteristics of adipose tissue. On MRI, it was hyperintense on both T1- and T2-weighted sequences and reduced signal intensity in FAT SAT sequences. In light of the imaging features and predominant adipose tissue component, the mass was diagnosed as a mediastinal lipoblastoma, and this diagnosis was confirmed by surgical histology.

Sommario Il lipoblastoma è una rara neoplasia mesenchimale, benigna, contenente tessuto adiposo, che colpisce l'infanzia e la prima adolescenza e che si localizza preferenzialmente nelle parti molli delle estremità. Nel nostro caso una bambina di 22 mesi è giunta alla nostra osservazione per una tumefazione della fossetta sovraclaveare che si rendeva manifesta durante il pianto. L'esame ecografico ha evidenziato una neoformazione del mediastino anterosuperiore a parziale sviluppo nella fossetta sovraclaveare destra, che appariva tenuemente iperecogena a margini netti. Tale neoformazione non presentava caratteristiche infiltrative nei confronti delle strutture limitrofe. La massa appariva alla TC disomogeneamente ipodensa, a densitometria adiposa, mentre alla RM risultava iperintensa nelle immagini T1 e T2-pesate, con abbattimento del segnale nelle sequenze FAT SAT. Le caratteristiche d'imaging ed in particolare la prevalenza di tessuto adiposo ci hanno suggerito l'ipotesi diagnostica di un lipoblastoma mediastinico, che è stata confermata dall'esame istologico eseguito sul pezzo operatorio.

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Introduction

The term “lipoblastoma” was used for the first time in 1926 by Jaffe to describe an atypical adipose-tissue tumor of the groin.

Lipoblastoma is a rare benign mesenchymal neoplasm containing adipose tissue, which occurs during childhood and early adolescence. It is localized preferentially in the soft tissues of the extremities. Less common locations are the trunk, neck, retroperitoneum, mediastinum, and perineum. The tumor is often asymptomatic: any symptoms that are present are directly related to the location and size of the mass. In two-thirds of the cases, the lipoblastoma is well circumscribed and may be partially or completely encapsulated. In the remaining third, it presents in a diffuse form that tends to infiltrate deeper adjacent organs, like muscles, and to recur locally (lipoblastomatosis). Histologically, the tumors are composed of mono- or multivacuolated lipoblasts, stellate mesenchymal cells, a plexiform capillary network, a myxoid stroma, and mature adipocytes organized within the stroma and separated by fibrous septa.

Case report

In December 2008, a 22-month-old girl was referred to our staff for a sonographic examination of the neck. The infant presented with a swelling in the supraclavicular region that became evident only when the intrathoracic pressure increased (e.g., when she cried). The past medical history was unremarkable.

The ultrasound examination was done with a Hitachi H21 scanner and linear (8–12 MHz) and convex (5–8 MHz) transducers. Above the epiaortic vessels on the right, US revealed an inhomogeneously hyperechoic oval mass measuring 4 cm in diameter. Color Doppler revealed no evidence of vascularization (Fig. 1). Conventional radiographic studies were within normal limits.

Fig. 1 US: Weakly hyperechoic mass located above the epiaortic vessels with no signs of vascular-wall invasion.

The patient was then subjected to a more in-depth diagnostic work-up, which (thanks to scanner availability) began with 64-slice computed tomography (Philips). This examination revealed a well-encapsulated oval mass that was inhomogeneously hypodense with a few hyperdense off-shoots. Densitometric findings were indicative of adipose tissue. The mass originated in the right anterosuperior compartment of the mediastinum and filled most of the right supraclavicular fossa (Fig. 2). The mass, whose maximum diameter was 4 cm, did not appear to have infiltrated any of the surrounding structures. A large portion was in contact with the clavicle although the two were separated by a clear cleavage plane; the mass extended upward, producing posterior displacement of the neck vessels without causing hemodynamically significant reductions in their diameters. It also caused compression and displacement of the trachea.

This examination was followed by magnetic resonance imaging (MRI) with a General Electric Signa 1.5-T scanner (Milwaukee, WI) with acquisition of T1- and T2-weighted coronal and sagittal sequences, with and without fat suppression. Contrast enhancement was not used. This examination confirmed the morphological features of the mass seen on CT, revealing a lesion that was hyperintense in both T1- and T2-weighted images (Figs. 3 and 4) with reduced intensity in FAT SAT sequences.

Based on the results of the imaging studies (in particular the abundance of adipose tissue), the absence of infiltrative characteristics, and the age of the patient, the mass was thought to be a mediastinal lipoblastoma. It was removed surgically, and the clinical diagnosis was confirmed by the histological examination. Follow-up, which included ultrasoundography every 3 months, has shown no signs of local recurrence.

Informed consent was obtained from the patient for publication of the case report.

Fig. 2 CT: Inhomogeneously hypodense mass with densitometric features of adipose tissues, sharply defined margins. The mass displaces surrounding structures but there is no sign of invasion.

Fig. 3 MRI: Axial T1-weighted: Inhomogeneously hyperintense mass with signal characteristics typical of fat and an expanding growth pattern.

Fig. 4 MRI: Sagittal T2-weighted scan of the same expanding mass shown in Fig. 3.

Discussion

The literature contains very few case reports regarding mediastinal lipoblastomas (1 every 100 cases). The incidence of this tumor peaks around the age of 23 years and decreases sharply after the age of 35 [1–3].

Lipoblastomas include three main components: lipoblasts, myxoid stroma, and mature adipocytes separated by fibrous septa. The appearance of the tumor on imaging studies depends on the histological features and varies according to the amounts of adipose, fibrous, and myxoid tissues it contains. According to the literature, lipoblastomas in very young patients tend to be composed mainly of myxoid tissue, which produces a hypoechoic image on ultrasonography and hypodensity on CT. On MRI, the high water content of these tumor produces a hypointense image in T1-weighted images and hyperintensity in T2-weighted sequences.

In older children, the adipose-tissue component predominates, and the appearance of the tumor is therefore identical to that of a lipoma. On US, CT, and MRI, these lipoblastomas exhibit well-defined margins; lobulation and internal septation are also frequent. The adipose tissue appears weakly hyperechoic on US, hypodense on CT, and produces a signal identical to that of subcutaneous fat on MRI.

Lipoblastomas can be differentiated from liposarcomas on the basis of patient age. The latter tumors are extremely rare in children under 10 years of age. Only 2 of the 2500 cases reported by the Armed Forces Institute of Pathology involved this age group, and both were diagnosed after the second year of life [5]. Liposarcomas also exhibit infiltrative growth, and its structure is inhomogeneous with thickened septa and nodules of nonadipose tissue that represent over 25% of the total mass. They also appear highly vascularized on color Doppler, CT, and contrast-enhanced MRI. Recently, some investigators have suggested that lipoblastoma can also be identified on the basis of specific molecular and cytogenetic markers (rearrangement of chromosome 8) [4]. It also needs to be differentiated from hibernomas, rare tumors composed of fetal brown adipose tissue that are generally observed in the third or fourth decade of life. Therefore, a mediastinal tumor composed largely of adipose tissue in a child is almost certainly a lipoblastoma.

These lesions have no malignant potential.

In our patient, the sonographic examination revealed an expansive neoplasm, which was characterized on CT and MRI by features indicative of a lipoblastoma composed mainly of adipose rather than myxoid tissue. This pattern contrasts with reports in the literature, which indicate that in lipoblastomas in children under 10 years of age, the myxoid component is usually predominant.

Conflict of interest statement

The authors have no conflict of interest.

Supplementary material

Supplementary data associated with this article can be found in the online version, at [doi:10.1016/j.jus.2011.01.002](https://doi.org/10.1016/j.jus.2011.01.002).

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