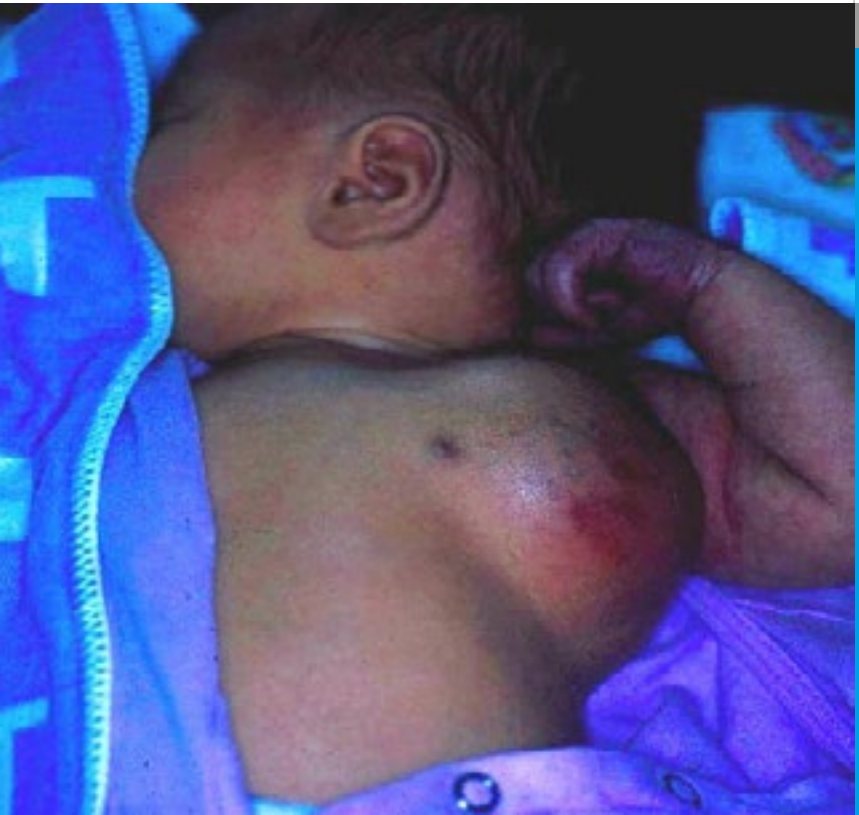
SWISS SOCIETY OF NEONATOLOGY

Congenital fibrosarcoma – a rare and curable tumor



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After an uneventful pregnancy, this 30-year-old G1/P1 presented herself in labor at 39 0/7 weeks of gestation. No prenatal care or ultrasounds had been obtained. A routine sonogram of the fetus on admission revealed a left-sided axillary mass of 5 cm in diameter. In anticipation of thoracopelvic disproportion, a Cesarean section was performed at 39 2/7 weeks of gestation. Arterial umbilical cord-pH was 7.28 and the infant adapted well with Apgar scores of 9, 10 and 10 at 1, 5 and 10 minutes, respectively .

Birth weight was 3500 g, birth length 49 cm and head circumference 34 cm. Physical examination was unremarkable except for a firm circumscribed seemingly encapsulated mass in the left axillary region, measuring approximately 5x5 cm. The tumor appeared well vascularized with regions of hemorrhage and probable necrosis. There were no palpable calcifications (Fig. 1).

Routine laboratory studies were normal. Alpha-fetoproteine: 30458 ug/l (normal for age), beta-HCG: 7.2U/l, neurone-specific enolase: 18.7 ug/l. On chest X-ray, there was a homogenous big soft tissue tumor without signs of calcifications; the left scapula was markedly detached from the thorax and there was no evidence of bone metastases or infiltrations (Fig. 2).

Sonographically there was a well-delineated round mass neighboring the thoracic cage which was not adherent to the underlying tissues with a mostly homoge-

nous ultrasonographic pattern of medium echogenicity (Fig. 3).

On MRI, a large left axillary oval mass (5x5 cm), situated between the pectoralis muscle and the thoracic wall, extending dorsally up to the scapula was demonstrated. The tumor was well delineated and mostly hyperintense on T2-weighted images and soft–tissue-isointense on T1-weighted images and showed noticeably inhomogeneous enhancement after intravenous contrast medium injection. The subclavian and axillary arteries were partially encased by the lesion. The brachial plexus could not be visualised. No enlarged regional lymph nodes were found (Fig. 4-7).

Surgery revealed an encapsulated tumour that was freed from the surrounding tissues. A completely occluded vein (probably the subclavian vein) led to a necrotic area, as already suspected on the MRI images (Fig. 8). The brachial plexus was completely embedded in the tumor. In order to free individual nerves and vessels the mass had to be cut in half. Residual tumor remained along the nerve-vessel-bundle (Fig. 9).

Histologically, the diagnosis of a congenital/infantile fibrosarcoma (T1b, N0, M0) was made: interlacing cords and sinuous bands of spindle cells with a high mitotic rate formed a typical "herringbone" pattern. Cells were immunoreactive to Vimentin, but not to CD 34, Desmin, Alpha SMA, S-100, CD 68 and MIB1 (Fig. 10).

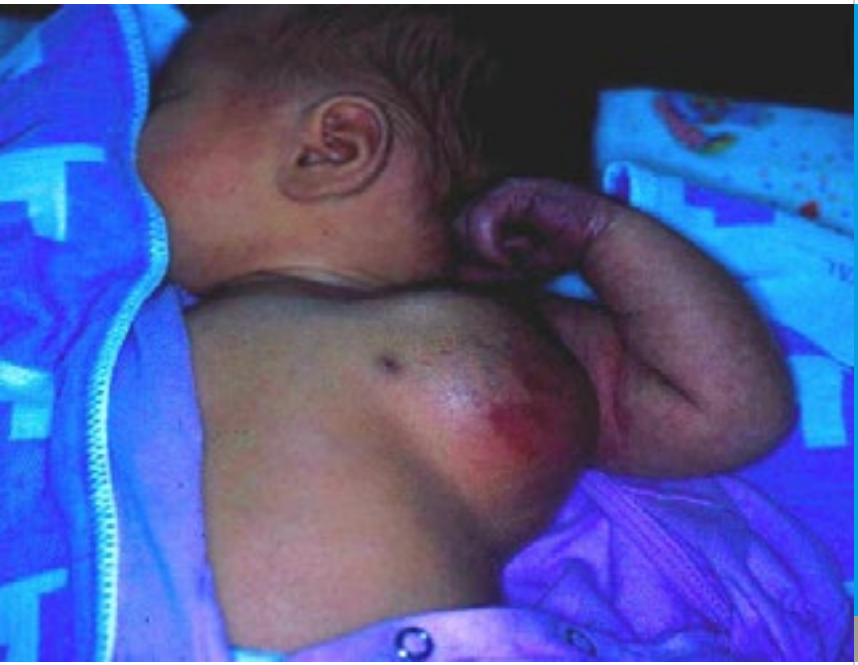

Fig. 1

Clinical appearance shortly after delivery.

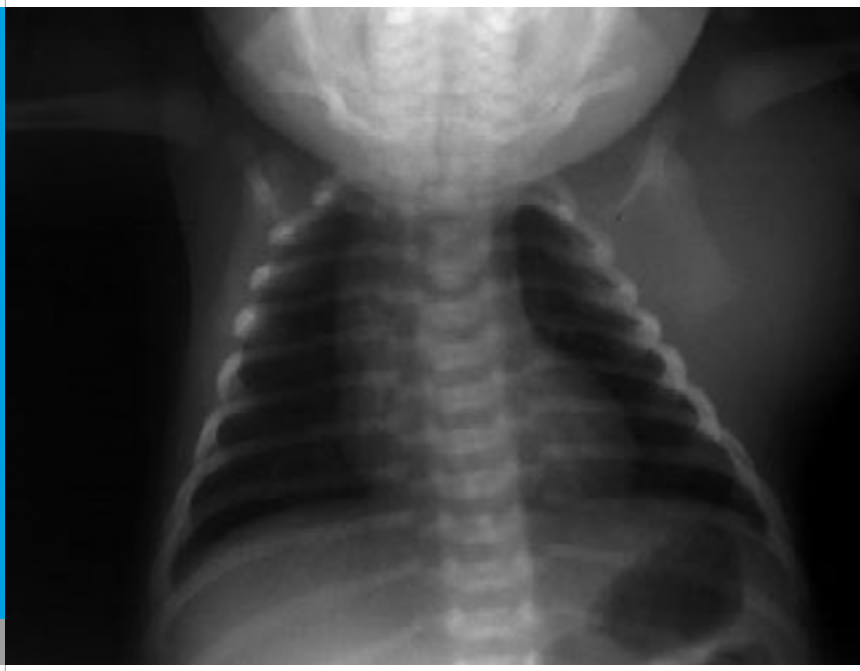

Fig. 2

Soft tissue mass displacing the left scapula.

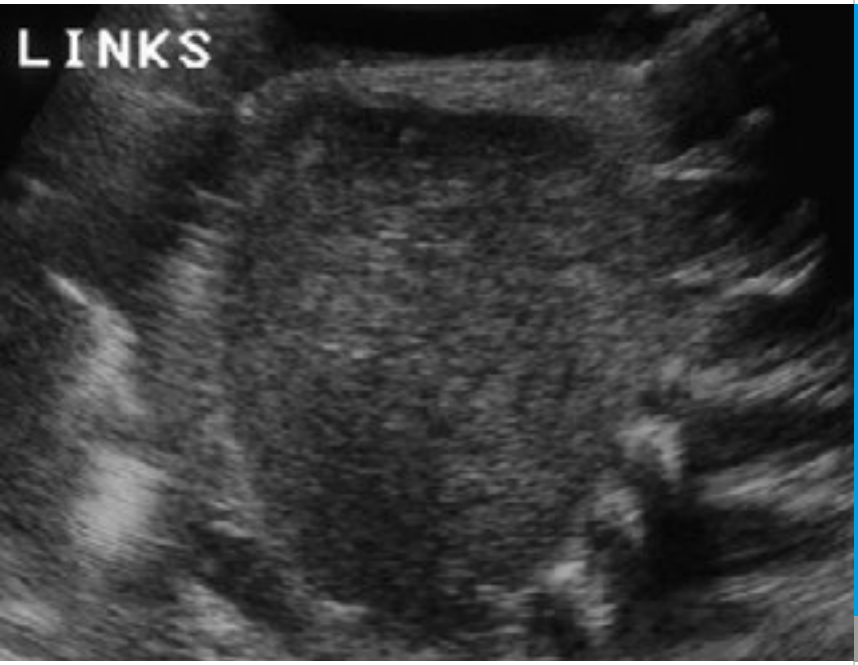

Fig. 3

Sonographic appearance of the tumor.

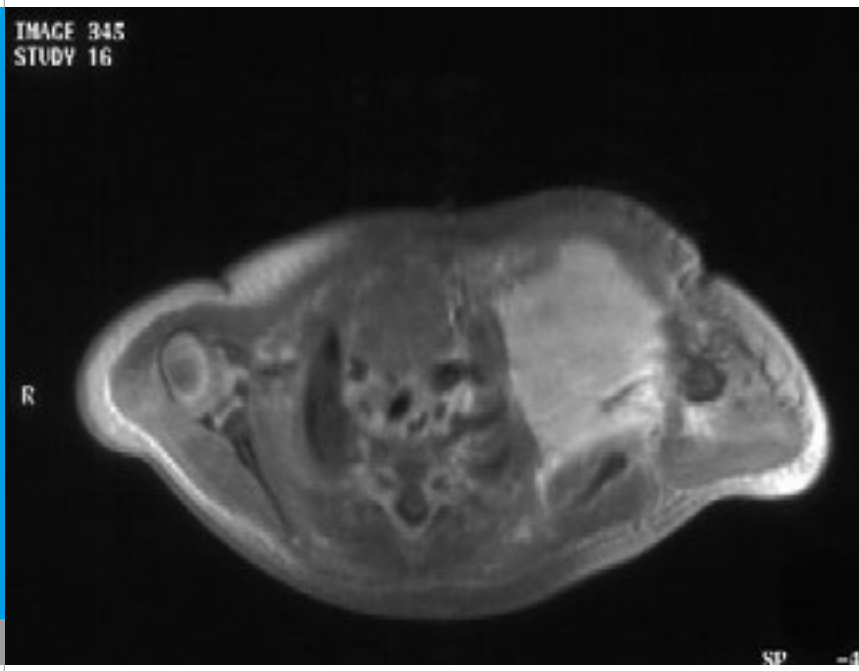

Fig. 4

MRI appearance of the soft tissue mass.

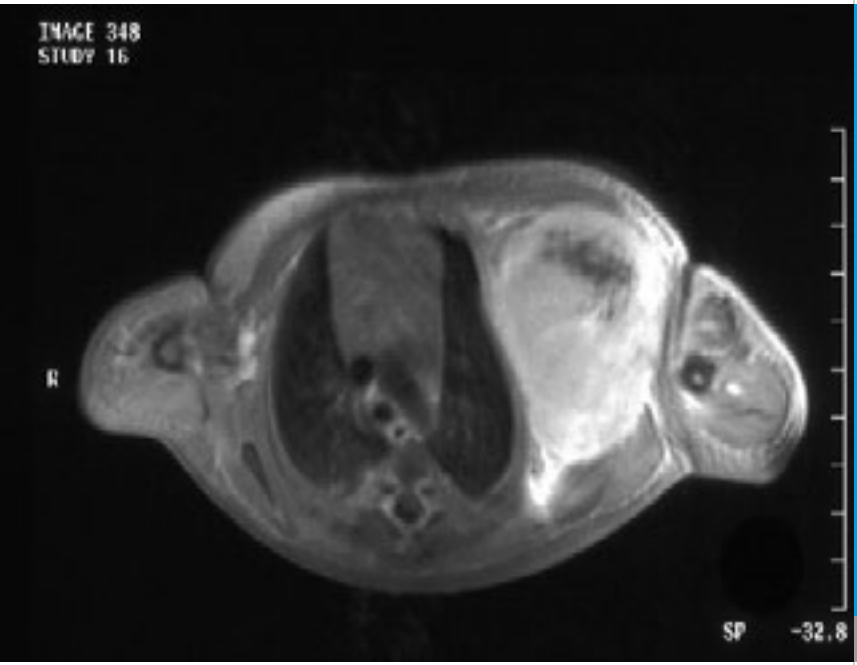

Fig. 5

MRI appearance of the soft tissue mass.

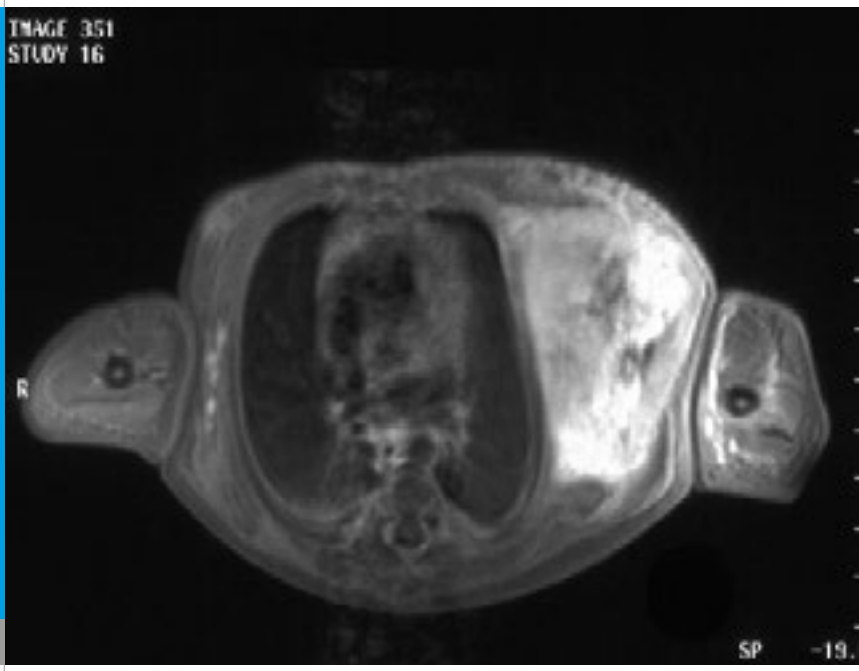

Fig. 6

MRI appearance of the soft tissue mass.

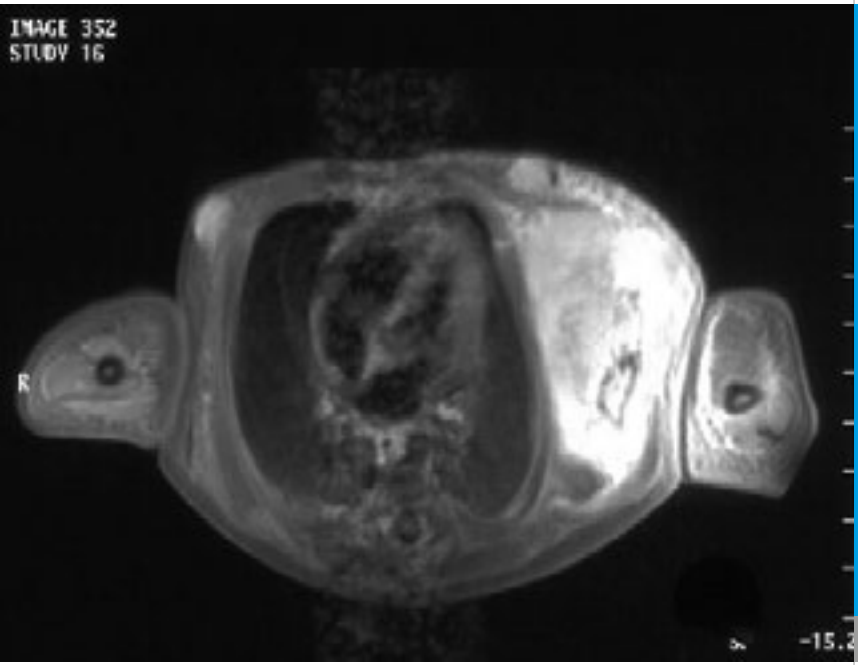

Fig. 7

MRI appearance of the soft tissue mass.

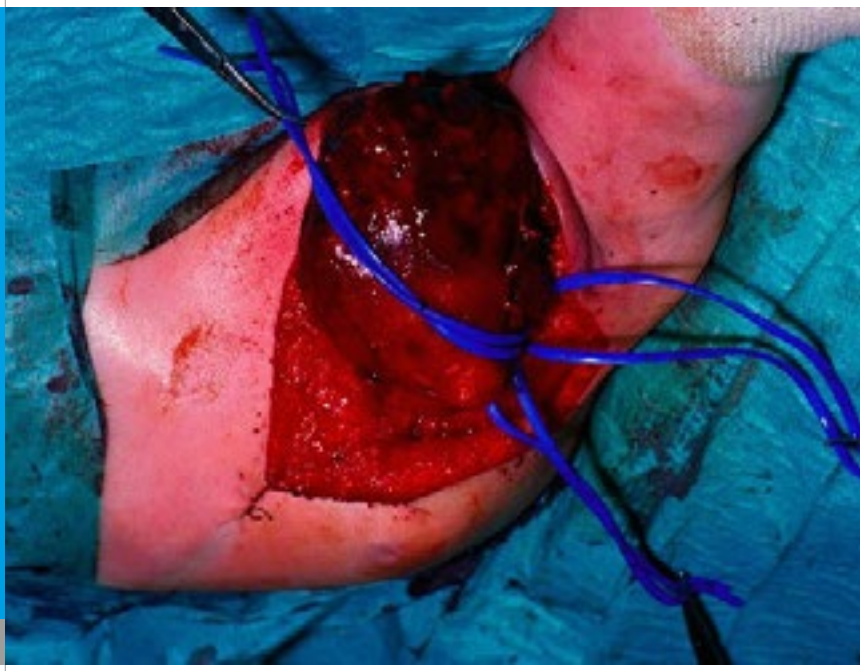

Fig. 8

Intraoperative appearance of the tumor at the time of resection.

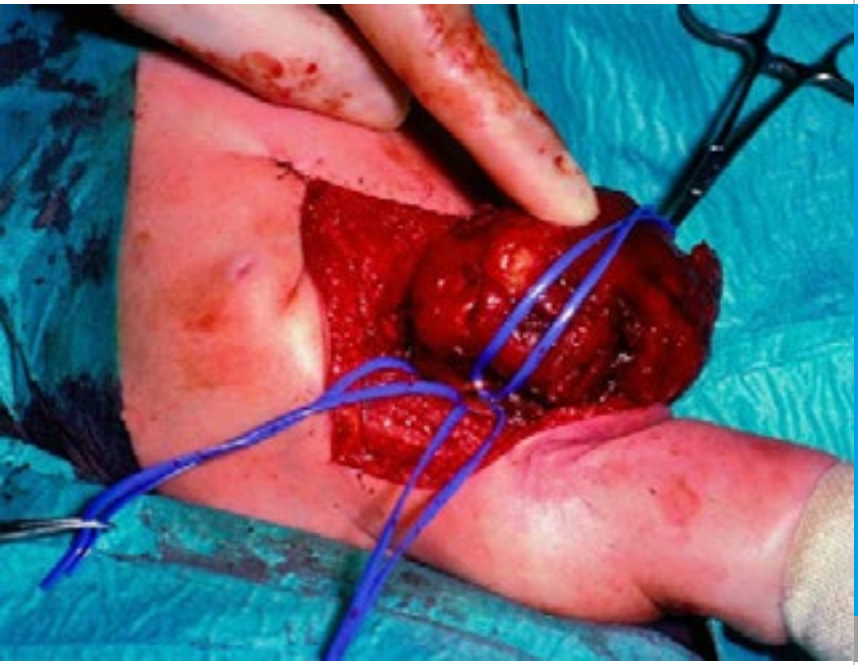

Fig. 9

Intraoperative appearance of the tumor at the time of resection.

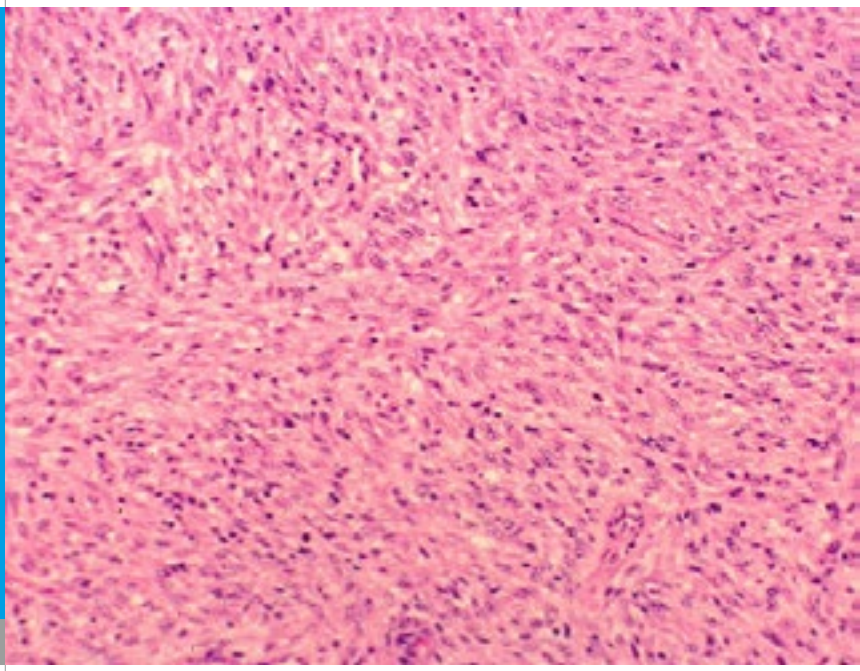

Fig. 10

Histology of resected tumor.

The differential diagnosis of congenital fibrosarcoma is extensive, and includes other benign and malignant spindle cell tumors of childhood, such as infantile myofibromatosis, benign and malignant nerve sheath tumors, smooth muscle cell tumors, rhabdomyosarcoma and inflammatory myofibroblastic tumor.

It is often impossible to make a clear distinction between infantile myofibromatosis and a fibrosarcoma in an infant with a cellular, mitotic active spindle cell neoplasm with no specific evidence of differentiation. In our case, the malignant neoplasm was well circumscribed. This, and its histological hallmarks of solid growth, dense cellularity, and prominent mitotic activity differentiate the tumor from infantile myofibromatosis.

Infantile fibrosarcoma is a rare soft tissue sarcoma predominantly occuring in children less than 24 months of age (1). The most recent Survey of Epidemiology and End Results program data gathered between 1975 and 1995 reported that the incidence of fibrosarcoma in infants was 5 per 1 million infants, almost equalling that of rhabdomyosarcoma, estimated to occur in 6 per 1 million infants (2). Despite histologic similarities, adult type fibrosarcoma is a clinically distinct entity with a poorer prognosis and a different cytogenetic profile (3).

Complete resection continues to be the treatment of choice; however, a majority of patients who have unresectable tumors at diagnosis show an excellent response to front-up chemotherapy (4-6).

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