Universitätsklinikum Hamburg-Eppendorf

AKK Altonaer Kinderkrankenhaus
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(Congenital) infantile fibrosarcoma - Rare diagnosis with good prognosis?

A German 3 year experience and literature review

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INTRODUCTION

Infantile fibrosarcoma is one of the most common non-rhabdomyosarcoma soft tissue sarcomas of childhood. It is often of benign nature, local recurrence after therapy occurs in 17-43% but metastases are uncommon (8%). More than one third of all cases are present at birth, therefore deserving the term „congenital fibrosarcoma". Optimal treatment consists of complete surgical resection. The role of chemotherapy seems limited and is today mainly regarded as an attempt to avoid extensive mutilating surgery .

AIM

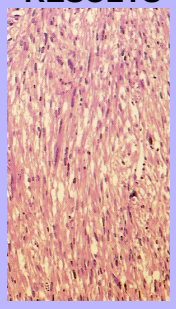
To review our limited experience and compare our results with the literature, with special emphasis on current treatment.

METHODS

A retrospective analysis of 3 infants treated for infantile fibrosarcoma (2007-2010).
Data were collected regarding the clinical course, treatment and outcome using patient files, operative reports and office notes.

A literature search was performed on MEDLINE using „infantile AND fibrosarcoma AND treatment AND prognosis" search terms, and compared with our treatment results.

RESULTS



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	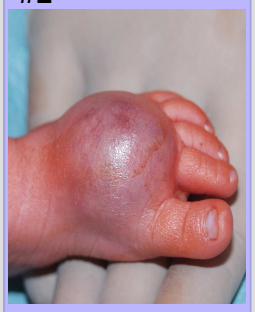	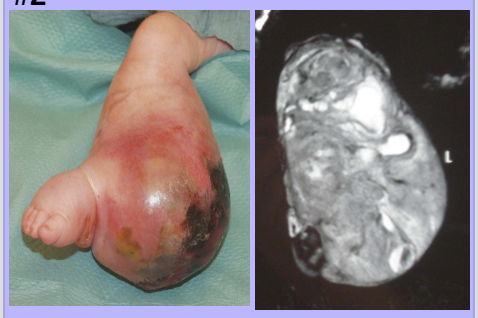	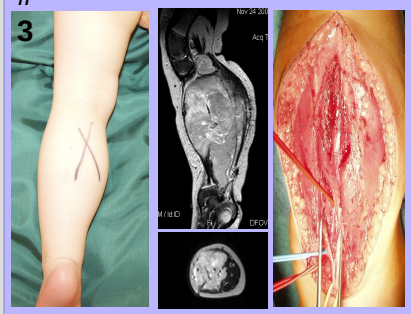
age diagnosis	19 d	13 d	7 months
1st symptoms	birth	birth	6 months
gender	F	M	M
tumor size	15 x 17 x 24 mm	12,5 x 8,0 x 9,5 cm	9,7 x 4,6 x 5,1 cm
treatment (CWS 2002) /	R1 resection	disarticulation of knee / R0 resection	neoadjuvant chemoTx VAC
R0 resection		(below knee prosthesis)	R2 resection / chemoTx VA /
tibial artery			incl. soleus muscle & posterior
EFS	41 months	40 months	4 months
clinical course requiring revision	uneventful	tumor bleeding & consumption thrombocytopenia	postoperative bleeding

LITERATURE REVIEW

Review of the literature shows that complete surgical resection is the corner stone of therapy. Tumor extension dictates whether extremity preservation is possible. The main new trend in treatment schemes is the use of up-front chemotherapy sometimes showing good tumor reduction which makes unresectable tumors resectable, leading to a significant decreased morbidity after radical surgery. ETV6-NTRK3 chimeric proteine – identified in both infantile fibrosarcoma and mesoblastic nephroma in 1998 – is regarded responsible for this sensitivity for chemotherapy.

CONCLUSION

The cases presented are conform with the literature concerning tumor localization - most frequenty in the lower extremities -, age at diagnosis, gender and prognosis. Survival rate was 100 % after a median follow-up of 28 months. Our study reflects that an excellent outcome is achievable when different treatment modalities are applied individually to each case – leading to good prognosis so far.

References - Himori et al 2004, Sandberg et al 2002, Chung et al 1976, Loh et al 2002, Kurkchubasche et al 2000, Ninane et al 1986, Bourgeois et al 2000, Knezevich et al 1998