OPEN ACCESS

Journal of Neonatal Surgery 2019; 8:13

CASE REPORT

Paraspinal Congenital Infantile Fibrosarcoma: A Case Report

Sivasankar Jayakumar,* Sanjana Venkateswaran, Sathish Manivel, Balamurali G

Department of Paediatric Surgery,Kauvery Hospital,TTK Road, Chennai 600028, India

ABSTRACT

Congenital infantile fibro-sarcomas (CIFS) are rare, locally aggressive mesenchymal tumors that usually develop from soft tissue of distal extremities. CIFS arising from paraspinal region are extremely rare and only few cases have been reported. A left thoracic paraspinal swelling in a female fetus was noted on 20-week antenatal scan of a primigravida. The swelling was monitored with regular antenatal scans. The baby was born at term and was well at birth. Serum AFP and Beta HCG were within normal age limits. MRI scan confirmed an aggressive tumor infiltrating the paraspinal muscles but not the spinal cord. The mass grew very rapidly in size within a few days, and was excised on day 7 of life. Histopathology and Immunohisto-chemistry diagnosed the mass to be a Fibrosarcoma. No chemotherapy or radiotherapy was given. The baby is well with no recurrence or residual deformity or neurological deficit after 18 months of follow up. Paraspinal CIFS are extremely rare but can be detected antenatally as early as 20 weeks of gestation. Complete surgical excision is the treatment of choice.

Key words: Infantile; Fibrosarcoma; Paraspinal; Congenital; Tumor; Neonate

INTRODUCTION

CIFS was first recognized in 1962.[1] This tumor arises from mesenchymal cells and composed of malignant fibroblasts within a collagen background. Fibrosarcomas account for 5%-10% of all soft-tissue sarcomas in infants. Approximately 40% of these lesions are present at birth with male preponderance and more than 80% are diagnosed within 1 year of age.[2] So far 82 cases of CIFS have been reported in the literature. CIFS are usually regarded as a tumor of borderline or low malignant potential with survival rates of 80-90%.[3] Ainsworth et al, reported three cases of CIFS involving the thoraco-lumbar paraspinal region.[4] Herein, we report a case of CIFS arising from the thoracic paraspinal region.

CASE REPORT

A large left thoracic paraspinal swelling in a female fetus was noted on routine 20-week antenatal scan of a primigravida. Fetal MRI showed no infiltration of the spinal cord. The swelling was monitored with regular antenatal scans but no significant growth was noted. Fetal biopsy was discussed with both parents for antenatal diagnosis of the lesion but declined. Despite, knowing the malignant potential, the mother decided to continue her pregnancy. The intrauterine growth of the fetus remained unimpaired and the baby was born at 37 weeks of gestation by elective c-section. Clinically the baby was well at birth and required no resuscitation or support. The paraspinal swelling was firm to hard and measured about 5x10x10cms.The baby was nursed in lateral and prone positions. Feeds were commenced immediately. However the hard protruding mass grew rapidly in size (10x10x10cms) within a few days (Fig.1a,1b) after birth.

Serum AFP (5753 ng/ml) and Beta HCG (< 1.2 mIU/ml) levels were within normal age limits. Ultrasound scan was initially done showed soft tissue swelling but the margins could not be delineated clearly. MRI confirmed an aggressive tumor, infiltrating the paraspinal muscles but not the spinal cord (Fig.2a,2b). After adequate counselling of parents, in view of rapid growth of tumor along with the potential risk of bursting and subsequent bleeding, surgical excision was done on day 7 of life without any prior biopsy. Intra-operatively, the tu-

Correspondence*: Sivasankar Jayakumar,Department of Paediatric Surgery, Kauvery Hospital, TTK Road, Chennai
E mail: shivajayakumar@gmail.com

Submitted: 10-10-2018 Accepted: 03-04-2019
Conflict of interest: Nil Source of Support: Nil

mor had a clear demarcation border to the surrounding tissues except anteriorly to the paraspinal muscles. Maximum excision was done without damaging the paraspinal muscles and radical dissection was not done. Post-procedure, the baby recovered and started tolerating oral feeds. The wound appeared healing well, but for a superficial wound dehiscence of 2cms at the paraspinal site.

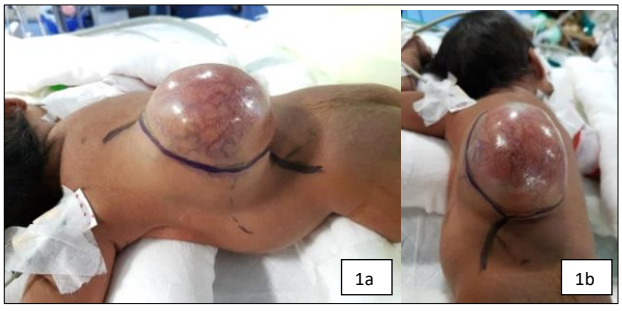

Figure 1: a) Lateral & b) AP view of mass of patient.

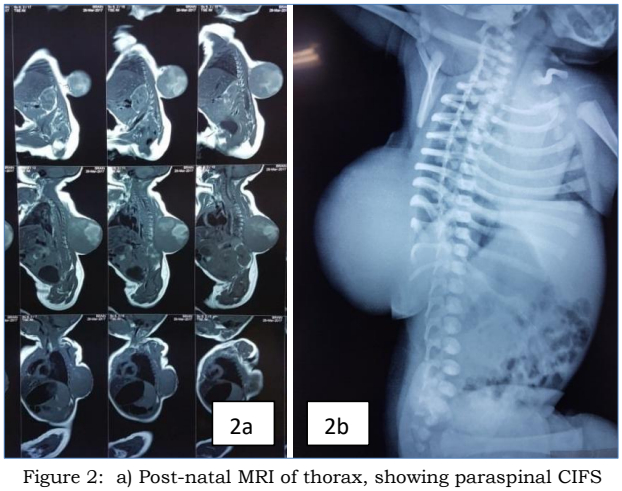

Figure 2: a) Post-natal MRI of thorax, showing paraspinal CIFS without any spinal cord infiltration b) Lateral plain x-ray of patient.

Histology showed fascicles of round to spindle cells with hemangiopericytoma patterns with mitotic figures of 4-6 /HPF (Fig. 3). Immunohistochemistry showed positivity for Vimentin; focal positivity to CD34 and SMA, and negative for S100, Desmin and Cytokeratin, suggestive of Infantile fibrosarcoma. Excision margins were complete on all sides except anteriorly where the paraspinal muscles where attached. Initially it was decided to observe, however as the wound dehiscence increased to more than 5cms with a large gap, a secondary wound closure was required. Along with a Z-plasty for the wound closure, a re-excision of residual tumor was attempted but limited as of risk of entering the intercostal space and further spillage of tumor into the pleural space. However, histopathological examination confirmed that the re-excision margins were adequate(R0). No adjuvant chemotherapy or radiotherapy was given throughout the stay. The baby recovered well and was discharged on day 21 of life. No evidence of recurrence has been noted clinically or on follow-up MRI scans so far. The baby remains well with no neurological deficit or recurrence at 18 months of follow up.

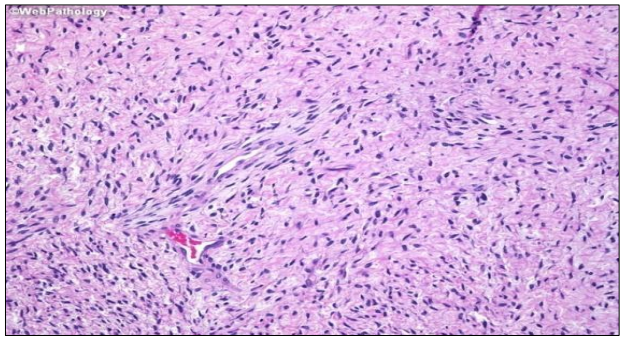

Figure 3: Histology of CIFS showing fascicles of round to spindle cells with hemangiopericytoma patterns. Mitosis of 4-6 /HPF.

DISCUSSION

About 40% of CIFS are diagnosed at birth and 60% before 3 months of age. Usually, antenatal detection and likely diagnosis is possible from 26 weeks of gestation on Ultrasound scan and MRI.[5] In our case, it was detected antenatally at 20 weeks of gestation. The three cases of CIFS reported in the thoraco-lumbar paraspinal region by Ainsworth [4] so far had presented between 1 week and 1 month of life but none of these cases were detected antenatally.

CIFS usually present as a solid mass at birth, and often the tumor is confused with hemangiomas or other vascular malformations. Histology of CIFS reveals a dense cellular proliferation of spindle, myxoid, small round cells with whorled patterns seen occasionally. Immunohistochemistry usually shows positivity for vimentin, as in our case. Frequently it also shows focal positivity with smooth muscle actin, desmin, cytokeratin and CD-34 but S-100 protein is negative.[6] Molecular biological analysis can be useful to identify the presence of the characteristic EVT6-NTRK3 translocation. This translocation is found in more than 70% of cases and is suggested to be an indicator of chemosensitivity by some authors.[7]

Surgery is the primary mode of treatment for CIFS. Excellent results are shown when complete resection with tumor free margins (R0) is achieved. In a study of 50 patients with CIFS, none of the R0 and R1 patients required chemotherapy. Overall the disease free rate was 84% after 3 years.[8] In our case, the tumor was easily amenable for complete excision as identified in the postnatal MRI. But, the initial surgical resection showed inadequate marginal excision on the anterior aspect. Initially it was decided to observe, however as the wound dehisced partially with a large gap requiring secondary surgical closure, re-excision of residual tumor was done

along with Z-plasty which achieved complete excision (R0) in our patient.

Pre-operative biopsy is helpful in establishing the diagnosis of a tumor prior to surgical excision as well as in choosing the most appropriate chemotherapy. Neo-adjuvant chemotherapy is used to shrink the tumor prior to resection as well as to treat local recurrences and metastases. However, chemotherapy in a newborn is challenging and always fraught with risks. The narrow therapeutic index of the chemotherapeutic agents and associated potential life threatening toxicities make it difficult to justify starting chemotherapy in newborns. If used, Actinomycin D and Vincristine are the most frequently used agents for CIFS.[8] However, when our patient was only a week old the tumor suddenly increased significantly in size within a few days posing high risk of imminent rupture. It was felt most appropriate to proceed with primary surgical excision and abandon biopsy as well as neo-adjuvant chemotherapy.

The three cases of CIFS in the thoraco-lumbar paraspinal region reported previously by Ainsworth et al received neo-adjuvant chemotherapy followed by surgery and were followed up for 39 (12 – 54) months without any recurrence. Although in our patient, neo-adjuvant chemotherapy or radiotherapy was not given; our patient is well with no recurrence or residual neurological deficit or deformity after 18 months of follow up. We believe as in our case and as previously reported [8], complete surgical excision (R0) alone is enough and chemotherapy may not be required in CIFS patients involving the paraspinal region without metastasis at presentation.

In cases with incomplete (microscopic) surgical excision (R1),conservatory surgical approach is still recommended. In the European cohort of 35 CIFS patients, only one local recurrence occurred out of 19 such patients, and was successfully treated with further surgery.[9] Nevertheless, when the recurrence occurs often a radical or mutilating surgery is needed and the overall consensus should be to try to avoid incomplete excision both macroscopic as well as microscopic. Although surgery is still seen as the cornerstone of treatment in this tumor, chemotherapy may play a critical role in large diffuse inoperable tumors as well as in local recurrence and in patients with macroscopic residual tumor.[9]

Conclusion

CIFS arising from the paraspinal region are extremely rare. Paraspinal CIFS can be detected antenatally as early as 20 weeks of gestation, as in our case. Our patient was managed with complete surgical excision (R0) alone and neo-adjuvant chemotherapy or radiotherapy was not required.

Consent: Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an under-standing that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

Author Contributions: All the authors contributed fully in concept, literature review, and drafting of the manuscript and approved the final version of this manuscript.

REFERENCES

1. Enos T, Hosler GA, Uddin N. Congenital infantile fibrosarcoma mimicking a cutaneous vascular lesion: a case report and review of the literature. J CutanPathol. 2017;44:193-200.
2. Kim HY, Cho YH, Byun SY, Park KH. A case of congenital infantile fibrosarcoma of sigmoid colon manifesting as pneumoperitoneum in a newborn. J Korean Med Sci. 2013;28:160-3.
3. Hayes-Jordan A. Recent advances in non-rhabdomyosarcoma soft-tissue sarcomas.SeminPediatr Surg. 2012;21:61-7.
4. Ainsworth KE, Chavhan GB, Gupta AA, Hopyan S, Taylor G. Congenital infantile fibrosarcoma: review of imaging features. PediatrRadiol. 2014; 44:1124.
5. Lagree M, Toutain F, Revillon Y, Gaussin G, Marie-Cardine A, Lemoine F, et al. Recurrent and metastatic infantile fibrosarcoma: A case report. Arch Pediatr 2011; 18:28-32.
6. Tarik E, lamiae R, Abdelouahed A, Tarik M, Hassan G, Anouar DM. Unusual case of congenital/infantile fibrosarcoma in a new born. Afr J Paediatr Surg. 2013;10:185-7.
7. Steelman C, Katzensteen H, Parham D, Stockwell C, Ricketts R, Abramowsky C, et al. Unusual presentation of congenital infantile fibrosarcoma in seven infants with molecular genetic analysis. Fetal PediatrPathol. 2011; 30:329-37.
8. Orbach D, Rey A, Cecchetto G, Oberlin O, Casanova M, Thebaud E,et al. Infantile fibrosarcoma: management based on the European experience. J Clin Oncol. 2010; 28:318-23.
9. Orbach D, Brennan B, De Paoli A, Gallego S, Mudry P, Francotte N, et al. Conservative strategy ininfantile fibrosarcoma is possible: The European paediatric Soft tissue sarcomaStudy Group experience. Eur J Cancer. 2016;57:1-9.