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Short Communication

Jejune and Crisscross-Infantile Fibrosarcoma

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Infantile fibrosarcoma is a paediatric mesenchymal neoplasm morphologically reminiscent of adult fibrosarcoma. Neoplasms such as congenital mesoblastic nephroma may be genetically associated with infantile fibrosarcoma and manifest concurrent chromosomal translocations. Up to 50% of infantile fibrosarcomas may reoccur although distant metastasis is exceptionally encountered. Disease associated survival ensues in a majority (> 90%) neoplasms.

Generally, infantile fibrosarcoma represents within paediatric population up to 10 years and is commonly discerned below 2 years of age. Tumefaction may ensue within axial regions and upper or lower extremities. Commonly, the neoplasm exemplifies significantly rapid tumour progression [1,2].

Majority (~70%) of infantile fibrosarcomas manifest with chromosomal translocation t (12;15) (p13; q26) which engenders ETV6-NTRK3 genetic fusion transcript as denominated with ETS variant gene 6 and neurotrophic tyrosine receptor kinase type 3. Aforesaid genetic transcript can be appropriately discerned with fluorescent in situ hybridization (FISH) or reverse transcriptase polymerase chain reaction (RT-PCR). Additionally, infantile fibrosarcoma delineates trisomy 8, 11, 17 or 20 [1,2].

Upon gross examination, neoplasm appears as an enlarged, firm to soft lesion wherein the superimposed cutaneous surface appears stretched, erythematous or ulcerated. Tumour magnitude may exceed > 30 centimetres. Cut surface appears as fleshy and grey/tan. Focal areas of myxoid alterations, cystic degeneration, haemorrhage or necrosis may be enunciated [2,3].

Upon microscopy, an inadequately circumscribed, lobulated tumefaction is discerned. Morphologically, tumour appears reminiscent of adult fibrosarcoma. The significantly cellular neoplasm is composed of miniature to enlarged, spindle shaped cells demonstrating cellular and nuclear atypia and pleomorphism [2,3].

Architecturally, neoplastic cells configure fascicles, a distinct herringbone pattern or prominent haemangiopericytoma-like areas with predominant staghorn vasculature. Mitotic figures are significant [2,3].

Focal necrosis and haemorrhage may ensue. Foci of dystrophic calcification or extramedullary haematopoiesis may be encountered. Tumefaction appears to infiltrate adjacent soft tissue with an 'irregular' perimeter of neoplastic cells [2,3].

Upon ultrastructural examination, tumour cells exhibit features of fibroblasts and myofibroblasts [3,4].

TNM staging of soft tissue sarcoma confined to trunk and extremities as per American Joint Committee on Cancer (AJCC) 8th edition [3,4].

Primary tumour

•TX: Tumour cannot be assessed •T0: No evidence of primary tumour •T1: Tumour <5 centimetres in greatest dimension •T2: Tumour ≥5 centimetres and <10 centimetres in greatest dimension •T3: Tumour ≥10 centimetres and < 15 centimetres in greatest dimension •T4: Tumour > 15 centimetres in greatest dimension.

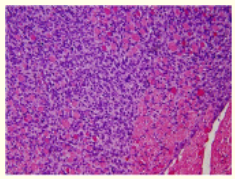

Figure 1: Infantile fibrosarcoma demonstrating fascicles of spherical and spindle shaped, fibroblastic cells with cellular and nuclear atypia with pleomorphism. The cellular component is intermingled with foci of haemorrhage and necrosis. Mitotic activity is discerned [7].

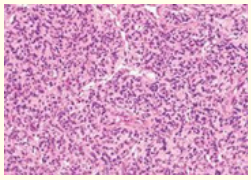

Figure 2: Infantile fibrosarcoma delineating bundles of spherical and spindle shaped, fibroblastic cells with cellular and nuclear atypia and pleomorphism. Mitotic figures are observed. Red cell extravasation is minimal [8].

Regional lymph nodes

•NX: Regional lymph nodes cannot be assessed •N0: Regional lymph node metastasis absent •N1: Regional lymph node metastasis present

Distant metastasis

•M0: Distant metastasis absent •M1: Distant metastasis present into sites such as pulmonary parenchyma

Histologic grade of soft tissue sarcoma of trunk and extremities [3,4].

•GX: Tumour grade cannot be assessed •G1: Tumour differentiation, mitotic count and necrosis quantified between total score of 2 and 3 •G2: Tumour differentiation, mitotic count and necrosis quantified between total score of 4 and 5 •G3: Tumour differentiation, mitotic count and necrosis quantified between total score of 6,7 or 8.

Tumour stages of soft tissue sarcoma of trunk or extremities [3,4].

•Stage IA: T1, N0, M0, GX or G1 •stage IB: T2, T3, T4, N0, M0, GX or G1 •stage II: T1, N0, M0, G2 or G3 •stage IIIA: T2,N0, M0, G2 or G3 •stage IIIB:T3, T4, N0, M0, G2 or G3 •stage IV: Any T, N1, M0, any G OR any T, any N, M1, any G.

Infantile fibrosarcoma is immune reactive to vimentin and exhibits a focal, variable immune reactivity to smooth muscle actin (SMA), desmin, S100 protein or CD34 [5,6].

Infantile fibrosarcoma requires segregation from neoplasms such as adult subtype of fibrosarcoma, infantile fibromatosis or myofibromatosis [5,6]. Infantile fibrosarcoma is appropriately treated with cogent surgical extermination of the neoplasm. Besides, adjuvant chemotherapy can be beneficially adopted with curative intent. Infantile fibrosarcoma is associated with superior prognostic outcomes, in contrast to adult fibrosarcoma [5,6].

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7. Image 1 Courtesy: Journal of perinatology
8. Image 2 Courtesy: Paediatric orthopaedic pathology