A 1-year-old male presented with a painless subcutaneous nodule in the left cervical region since birth [Figure 1]. On physical examination, the nodule was 2 cm in diameter, mobile, and nontender. There was no other lymphadenopathy or any organomegaly. Ultrasonography revealed a hyperechoic soft-tissue mass measuring 2 cm × 2 cm × 1 cm. A diagnosis of benign spindle cell lesion was made on fine-needle aspiration cytology and excision was advised.

Local excision of the mass was done. Grossly, it was firm to soft with gray-white and gray-yellow areas. The histopathological section examined showed immature round mesenchymal cells, islands of mature adipose tissue, and spindle-shaped fibroblastic cells in a collagenous stroma [Figure 2]. No mitotic figures, necrosis, or atypia was identified. On immunohistochemistry (IHC), vimentin was positive in the spindle cells and in the mesenchymal cells [Figure 3a]. CD34 showed immunoreactivity in the mesenchymal cells [Figure 3b].

**Question**

What is Your Diagnosis?
Answer
Fibrous hamartoma of infancy.

Discussion
Enzinger suggested the term fibrous hamartoma of infancy to emphasize its organoid microscopic appearance and its frequent occurrence at birth and during the immediate postnatal period.\[1\] It is rare in children >2 years of age and is more common in boys with a male to female ratio of 2:1.\[2\] The common sites of involvement are the axillary region, upper arm, upper trunk, inguinal region, and external genital areas.\[1,2\]

Cytological findings of fibrous hamartoma of infancy include adipose tissue fragments and fibroblastic cells in clusters as well as lying discretely associated with the myxoid and collagenous matrix.\[3\] Fibrous hamartoma of infancy is histologically characterized by varying proportions of tissues arranged in an organoid pattern comprised of fibrous trabeculae or septae composed of spindle cells separated by collagen, myxoid foci-containing primitive round or stellate mesenchymal cells and interspersed mature fat.\[2,4\] Morphologically, it has a varied differential diagnosis. Infantile fibromatosis arises primarily in muscle rather than in the subcutis, locally aggressive containing both myofibroblastic and adipocytic areas, but lacks the primitive mesenchymal cells seen in fibrous hamartoma.\[4\] Calcifying aponeurotic fibroma occurs in more distal locations such as the palm of older children and shows distinctive spotty calcifications surrounded by radiating fibroblasts and osteoclast-like giant cells.\[3\] The characteristic organoid pattern helps to distinguish fibrous hamartoma of infancy from rhabdomyosarcoma.\[6\]

Immunohistochemically, vimentin is reactive in the spindle cells and primitive mesenchymal cells, while S100 immunoreactivity is present only in the mature adipose tissue.\[2,4\] CD34 is positive in the mesenchymal cells, whereas smooth muscle actin is positive in the spindle-shaped fibroblastic cells.\[2,4\]

Clinically, it is often misdiagnosed as lipoma, dermatofibroma, neurofibroma, and hemangioma or as an enlarged lymph node.\[6\] Thus, characteristic histomorphology along with immunohistochemistry helps in arriving at a final diagnosis.

Simple excision is curative, and incomplete excision does not seem to carry a significant risk for recurrence.\[2,5\] However, 16% of cases recur locally within a few months after primary excision due to incomplete excision but are nondestructive and cured by re-excision.\[1,2,5\]

Learning points
- Fibrous hamartoma of infancy is a rare benign tumor
- It frequently occurs at birth and during the immediate postnatal period
- Histopathologically it is characterized by a triphasic organoid pattern
- Clinically, it is often misdiagnosed as lipoma, dermatofibroma, neurofibroma, and hemangioma or as enlarged lymph node
- It has an excellent prognosis with local excision being curative.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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