Intradermal spindle cell/pleomorphic lipoma: Case report and review of the literature

Sir,

Spindle cell and pleomorphic lipoma represent the same lipoma entity. The lipoma usually arises from the subcutis and rarely localizes in the dermis, leading to misdiagnosis. We found only five papers published in English literature that has addressed this exceedingly rare benign lipogenic tumor.1–5

A 60-year-old man presented with a 1-year history of a lump on the right lateral aspect of his nose. There was no history of trauma or previous skin diseases. Family and personal history were unremarkable. Physical examination revealed a red-colored, soft, well-demarcated papule 5 mm in diameter on the right lateral aspect of the nose [Figure 1]. The regional lymph nodes were not palpable. The lesion was removed with excision biopsy under local anesthesia.

Histological examination revealed a circumscribed dermal neoplasm without any capsule [Figure 2], which was filled with abundant mature adipose tissue in the central area and ovoid spindle cells confined to the peritumoral margin [Figure 3a]. The lipomatous area was consisted of many mature adipocyte cells, and some small mono-, bi-, or multivacuolated lipoblasts with atypical nuclei. In addition, floret-like bizarre and multinucleated giant cells were intermingled with the spindle-celled and adipocytic components [Figure 3b]. Immunohistochemical studies demonstrated that spindle-shaped cells were positive for CD34 [Figure 4] and were negative for S100 protein. Diagnosis of intradermal spindle cell and pleomorphic lipoma was made; continuous follow-up of the patient for 12 months revealed no recurrence.

Cutaneous spindle cell and pleomorphic lipoma is an extraordinarily rare benign adipose tissue tumor, representing approximately 9.8% of all spindle cell lipomas and 1% of tumors with adipocytic differentiation.1 There have been a total of 38 cases reported in the literature1–5 since it was initially described by Nigro in 1987.3 The tumor has shown a predilection for males, and has occurred...
Letters to the Editor

Recurrences

Benign

First Table

Predominance

Character

References

Immunohistochemical analysis of CD34 expression by spindle cell/pleomorphic lipoma and well-differentiated liposarcoma

Table 1: Clinical differences between spindle cell/pleomorphic lipoma and well differentiated liposarcoma

| Disease                          | First described | Areas affected                                      | Gender               | Predominance | Recurrences | Character          | Histopathology                                                                 | Immunohistochemistry                          | Cyto genetic findings |
|----------------------------------|-----------------|----------------------------------------------------|----------------------|--------------|--------------|-------------------|--------------------------------------------------------------------------------|-----------------------------------------------|----------------------|
| Spindle cell lipoma              | 1975, Enzinger and Harvey | Shoulders, backs, and neck (classically) | Male                  | Middle-aged to elder | Uncommon     | Benign            | Including a variable mixture of short fascicles of minimally atypical spindle cells with hyperchromatic nuclei, mature fat cells, collagenous to myxoid stroma with high numbers of mast cells and bright eosinophilic, ropcy collagen bundles | Nuclear and cytoplasmic reactivity for S100 (adipocytic cells) and cytoplasmic positivity for vimentin. Immunoreactive for vimentin and CD34 (spindle cells) | Presence of the same cytogenetic findings, such as monosomy or partial loss of 16q, and unbalanced aberrations of 13q |
| Pleomorphic lipoma               | 1981, Shmookler and Enzinger | Head/neck, trunk, breast, thigh, buttock, ears, limbs, vulga | A peculiar admixture of variably sized fact cells, bizarre, pleomorphic, floret, multinucleated giant cells, and occasional multivacuolated lipoblasts | | | | | |
| Well differentiated liposarcoma   | 1994, Dei Tos AP and Mentzel T | Subcutaneous tissue of the extremities, the trunk, and the head and neck region | Female               | 40-60, middle aged | Often recur locally, but rarely develop distant metastasis | Atypical/low-grade malignant | Atypical lipogenic neoplasm composed of atypical adipocytes showing striking variation in size and shape with scattered enlarged and hyperchromatic nuclei associated with slightly atypical spindle-shaped neoplastic cells | Atypical spindled tumor cells often staining positively for CD34 in many cases and lack immunohistochemically detectable expression of MDM2 and/or CDK4 in most cases | Absence of q13-15 regions of chromosome 12 and a monosomy of chromosome deletion of material of the long arm of chromosome 13 |

Previous studies demonstrated loss of 13q in a group of morphologically similar entities including cellular angiofibroma, mammary-type myofibroblastoma, and spindle cell lipoma. In addition, relatively recent studies by Chen et al. confirmed loss of Rb protein expression among this group of tumors and reinforced their pathogenetic relationship.

Differential diagnosis is extensive and includes both benign and malignant soft tissue tumors, especially well-differentiated spindle cell liposarcoma [Table 1]. Clinically, well-differentiated spindle cell liposarcoma presents with a locally aggressive growth and may recur, whereas metastases do not occur. Histologically, the spindle cells contain slightly enlarged, fusiform nuclei that are sometimes hyperchromatic and irregularly shaped. These are set in a collagenous stroma that may show hyalinization or myxoid changes. Immunohistochemically, an expression of CD34 by the spindle cells has been reported in many cases and lacks amplification of MDM2 and/or CDK4 in most of the cases analyzed. For curative therapy of liposarcoma, surgical excision remains the mainstay of management. A wide surgical margin is important to prevent local recurrence with or without additional postoperative radiotherapy and/or chemotherapy. Well-differentiated liposarcoma rarely develop distant metastasis but often recur locally.

Over a wide range of ages (from 20 to 85 years) with a mean age of 42 years. It usually presents as a slow-growing cutaneous nodule measuring less than 2.5 cm. The presenting symptoms and clinical signs of our patient were similar to previously described cases. It can occur in the head/neck region, trunk, breast, thigh, buttock, ear, lower limbs, upper limbs, and the genital region. Grossly, intradermal spindle cell and pleomorphic lipomas are ill defined and often unencapsulated. Microscopically, it is characterized by the presence of pleomorphic, bizarre, spindle cells, and floret-type giant cells. Among the 38 cases reported, 15 cases merely consisted of a spindle cell component. One report of pure cutaneous pleomorphic lipoma localized to the nasolabial region of a 59-year-old man is described.

Spindle cell lipoma localized to the ear, lower limbs, upper limbs, and the genital region.

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Among the reported cases, only one patient, a 71-year-old male, developed local recurrence at the site of a previous excision 21 years later. Complete gross excision with complete histopathologic margin evaluation achieves an almost complete cure rate. No case with metastasis or disease-related death has been reported.

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4. Chen et al. Immunohistochemical analysis of CD34 expression by spindle cell/pleomorphic lipoma and well-differentiated liposarcoma.

5. Addition, relatively recent studies by Chen et al. confirmed loss of Rb protein expression among this group of tumors and reinforced their pathogenetic relationship.
Spindle cell/pleomorphic lipoma generally presents as a solitary, soft, and slowly enlarging mass, with a variable size measuring 1–13 cm. This is a type of benign lipogenic tumor composed of primitive CD34-positive spindle cells, floret-like multinucleated giant cells, and mature adipocytes. Cytogenetically, most spindle cell/pleomorphic lipomas show 16q or 13q abnormalities. The treatment of choice is complete local excision, and recurrence is extremely rare. In our case, the lesion lasted for one year without aggressive and invasive growth. Histologically, there were no atypical lipogenic cells. Spindle-shaped neoplastic cells were characterized by slightly enlarged, fusiform nuclei that were sometimes hyperchromatic and irregularly shaped. The patient was discharged and has remained disease free for a follow-up period of 8 months. Thus, a diagnosis of intradermal spindle cell/pleomorphic lipoma was made. Other disorders that may be misdiagnosed includes solitary fibrous tumor, cutaneous neurofibroma, cutaneous angiomyolipomas, and lipoblastoma [Table 2].

Microscopically, in contrast to a majority of reported cases, our case was well circumscribed and entirely intradermal. Therefore, this case represents the 39th report of intradermal spindle cell and pleomorphic lipoma in the English literature.

S-100 is usually positive in adipocytes of spindle cell lipoma. In our case, S-100 was positive in fat cells and negative in spindle-shaped cells.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his
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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Facial multiple xanthogranuloma in an adult

Sir,

Xanthogranuloma is a normolipemic non-Langerhans-cell histiocytosis. Though this condition predominantly occurs in infancy and childhood and is also called "juvenile xanthogranuloma", around 10% of all affected patients are adult. Solitary lesions have been observed in approximately 90% of either juvenile or adult cases.1,2

The appearance of multiple lesions is rare in the juvenile form of xanthogranuloma and even rarer in the adult form. Here, we report a case with multiple adult xanthogranulomas who presented with brown, flat wart-like papules on the face.

A 23-year-old man with a 5-year history of brown papules on the face was referred to our dermatology department. Four years ago, he was diagnosed with flat warts and received laser treatments at a local clinic with no response. On examination, there were numerous small light brown papules distributed over the patient's cheeks, root of the nose, temples, and forehead [Figure 1a and b]. These papules were ill-defined and coalescent, round or irregularly shaped with a shiny surface and size of 0.2 cm to 0.5 cm. [Figure 1c]. These lesions were soft upon palpation and did not exhibit any subjective symptoms. No similar lesion was found on any other part of the patient's body. Ophthalmologic examination of the patient was normal. The patient denied any special exposure, such as fatty food, outdoor work, drugs, radiation, chemicals and so on. No other family member was affected. The laboratory test results, including routine hematological examination, liver and renal function tests, and serum lipid and sugar levels were all normal. Chest and skull radiographs and abdominal ultrasonographic examination revealed no abnormalities. A biopsy was taken from the left temporal region which showed dermal infiltration with histiocytes, lymphocytes, foam cells, and eosinophils [Figure 2a]. Touton giant cells which are multinucleated giant histiocytes with wreath-shaped nuclei and foamy cytoplasm were also seen [Figure 2b]. The infiltrating histiocytes expressed CD68 [Figure 2c] and did not express either S-100 or CD1a, as determined by immunohistochemistry. A final diagnosis of multiple adult xanthogranuloma was made based on the previously mentioned clinical, histopathological, and immunohistochemical findings.

Xanthogranuloma is a benign, self-healing condition which is clinically characterized by one to a few orange or brown papules or nodules commonly located on the upper body, particularly the head and neck.2-4 The male-to-female ratio of its incidence is approximately 1.3:1.3,4 Approximately 20–30% of juvenile xanthogranuloma cases occur at birth with up to 80% of cases occurring in the first year of life.1 Only around 10% of xanthogranuloma cases manifest in adulthood, particularly between the late 20s and early 30s.5 Most of the adult patients have...