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 literature[1-5] since it was initially described by Nigro in 1987.[3] The tumor has shown a predilection for males, and has occurred

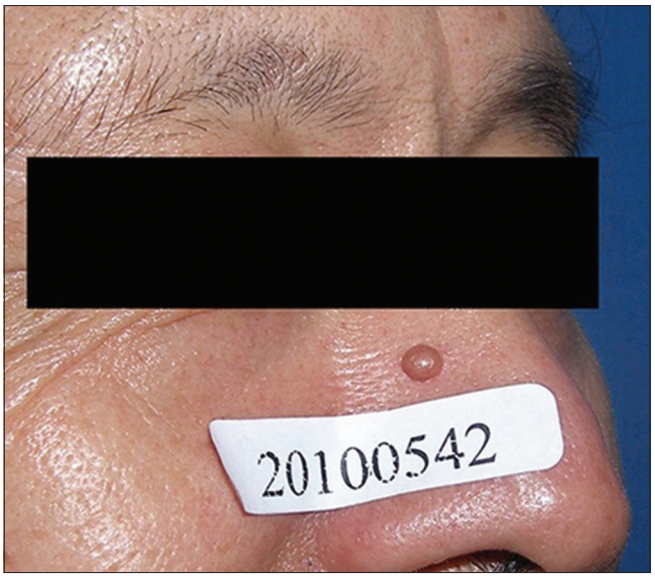

Figure 1: A single, red-colored, soft, well-demarcated nodule measuring 5 mm, present over the right lateral aspect of the nose

over a wide range of ages (from 20 to 85 years) with a mean age of 42 years.[1] It usually presents as a slow-growing cutaneous nodule measuring less than 2.5 cm.[1] 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.[2] 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 59-year-old man is there in the literature.[1-5] 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.

Previous studies demonstrated loss of 13q in a group of morphologically similar entities including cellular angiofibroma, mammary-type myofibroblastoma, and spindle cell lipoma.[4] 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.[5]

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.[9] 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.

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

Disease	First described	Areas affected	Gender	Predominance	Recurrences	Character	Histopathology	Immunohisto chemistry	Cytogenetic findings
Spindle cell lipoma[6]	1975, Enzinger and Harvey	Shoulders, backs, and neck (classically)	Male prepond erance	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, ropy 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[2]	1981, Shmookler and Enzinger	Head/neck, trunk, breast, thigh, buttock, ears, limbs, vulgar					A peculiar admixture of variably sized fact cells, bizarre, pleomorphic, floret, multinucleated giant cells, and occasional multivacuolated lipoblasts		
Well differentiated liposarcoma[7]	1994, Dei Tos AP and Mentzel T	Subcutaneous tissue of the extremities, the trunk, and the head and neck region	Female prepond erance	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 imunohisto chemically detectable expression of MDM2 and/or CDK4 in most cases	Absence q13-15 regions of chromosome 12 and a monosomy of chromosome deletion of material of the long arm of chromosome 13

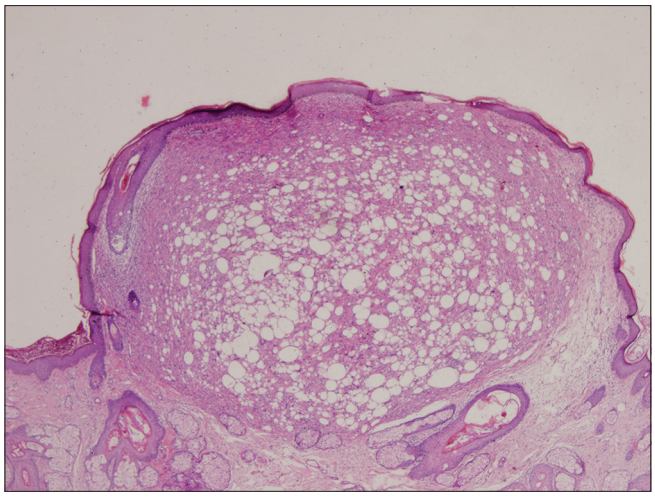

Figure 2: The neoplasm is well circumscribed and entirely intradermal, characterized by spindle cells confined to the peritumoral margin (H and E, ×100)

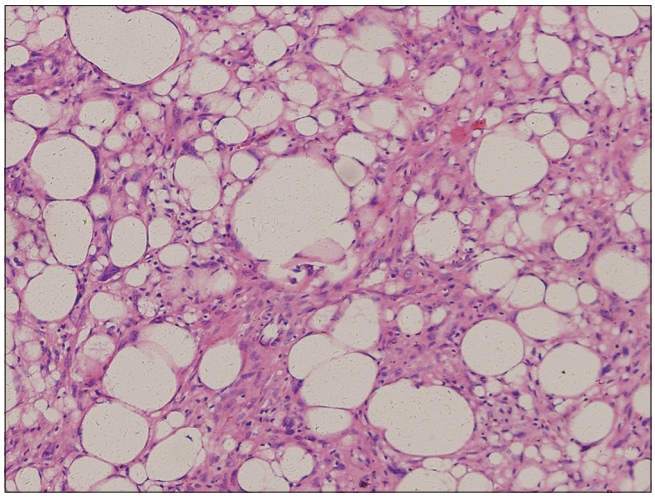

Figure 3a: The tumor was composed of abundant mature adipose tissue in the central portion and ovoid spindle cells confined to the peritumoral margin (×200)

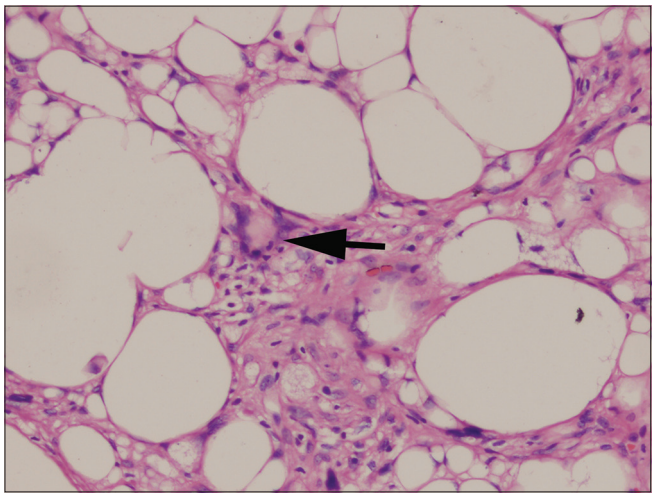

Figure 3b: The lipomatous area consists of an admixture of mature adipocytes, and small mono-, bi-, or multi vacuolated lipoblasts with atypical nuclei (H and E, ×200). Floret giant cells are highlighted by arrows

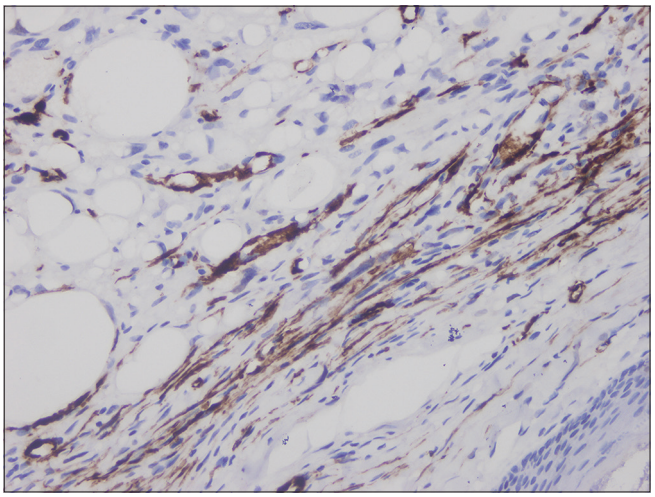

Figure 4: Immunohistochemical study shows $CD34^+$ spindle cells in spindle cell and pleomorphic lipoma (×400)

Spindle cell/pleomorphic lipoma generally presents as a solitary, soft, and slowly enlarging mass, with a variable size measuring 1–13 cm.[10] This is a type of benign lipogenic tumor composed of primitive CD34-positive spindle cells, floret-like multinucleated giant cells, and mature adipocytes.[2] Cytogenetically, most spindle cell/pleomorphic lipomas show 16q or 13q abnormalities.[11] 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

Table 2: Differential diagnoses based on clinical examination alone

Disease	Size (cm)	Location	Age	Gender	Border	Texture	Recurrence	Subjective symptoms
SC/PL	1-13	In the posterior head and neck, but also in the shoulder and back	Middle-aged	Male	Clear and slow growth	Soft	Very uncommon (benign)	No pain and pruritus
STFs[11]	1-17	Have been described in nearly every organ and site	Middle-aged adults between 20 and 70 years	No gender predilection	Clear and slow growth	Soft	Some aggressive (local recurrence/ metastasis, most benign)	Asymptomatic (some may feel pain)
CNF[12]	Any size and shape	Congenital disease involves nerve, muscle, bone, internal organs and skin	Mainly during the childhood and adolescence	Females	Clear, skin colored to yellowish papules	Soft	Some may be recurrent and malignant	Pain and functional loss when tumors press nerve
CAL[12]	1-4	Frequently located on the ear or acral skin	In the fifth or sixth decade of life	Males	Clear, raise, firm	Tender or hard	No recurrence	Not painful but patients may ache when the capsule connected with small nerve
LPB[13]	3-7	The extremities are the most common locations, but many other sites can be involved	In infants and eraly children, rarely in young adults	No gender predilection	Localized (LPB) or diffuse (lipoblastomatosis)	Soft	No recurrence generally (benign)	Painless nodule

SFT: Solitary fibrous tumor, CNF: Cutaneous neurofibroma, CAL: Cutaneous angiomyolipomas, LPB: Lipoblastoma, SC/PL: Spindle cell/pleomorphic lipoma

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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