Introduction
Indeterminate dendritic cell tumor (IDCT) is an extraordinarily rare disease with features common with Langerhans cell histiocytosis with respect to morphology and immunophenotype, but lacking Birbeck granules characteristic of Langerhans cells. Characterized by the proliferation of dendritic cells, it expresses CD1a and S-100 protein but lacks Langerin expression and Birbeck granules.[1] IDCT can occur as multiple solid red, yellow, or reddish-brown papulonodules, or, less commonly, as a solitary lesion.[2] It is almost always restricted to the skin without systemic symptoms.[3] We, herein, report a case of IDCT with the possibility of multiorgan involvement.

Case Report
A 23-year-old male patient presented to the Medicine department for difficulty in breathing and weight loss since 8 months. A dermatology referral was sought for skin lesions.

A general physical examination revealed pallor with enlarged submandibular and occipital lymph nodes.

A cutaneous examination showed multiple erythematous papules and nodules on the trunk, face and, the scalp [Figure 1a]. There was ulceration of both the axillae with scarring [Figure 1b and c]. Some lesions showed central umbilication and some were covered with adherent scales [Figure 1d]. Differential diagnosis of Cryptococcosis, Pyoderma gangrenosum, Hidradenitis suppurativa and, Langerhans cell histiocytosis was made.

Brachial breath sounds were heard over the lower chest bilaterally with occasional crepitations.

Peripheral blood smear showed normocytic normochromic red blood cells. There was neutrophilic leucocytosis with relative lymphopenia. Atypical cells or blasts were not seen. Hemoglobin was 8.5 gm/dL. Erythrocyte sedimentation rate was 37 mm/hour, C-reactive protein (CRP) was 10.2 mg/L, and the total serum protein was low (4.8 gms/dl). A high-resolution computed tomography (HRCT) of the thorax showed multiple bullae with panlobular emphysematous right residual lung with left-sided mild pleural effusion. An ultrasonography (USG) of the whole abdomen showed multiple complex cystic and solid lesions in the liver with mildly coarsened hepatic echotexture and minimal ascites. A non-contrast computed tomography (NCCT) of the brain and skull showed bony calvarium intact with no other abnormalities.

Fine needle aspiration cytology (FNAC) from the lymph node showed only reactive lymphoid cells. Biopsies from the trunk

Figure 1: (a) Ulceration and scarring of the right axilla. (b) Ulceration and scarring of the left axilla. (c) Some papules showed central umbilication (d) Multiple papules and nodules on the trunk.

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Figure 2: (a) Skin biopsy showing aggregates of tumor cells resembling Langerhans cells throughout dermis (H and E, 40X). (b) Tumor cells exhibiting varied shaped nuclei with abundant cytoplasm (H and E, 400X)

Figure 3: (a) Tumor cells positive for CD1a (DAB, Immunohistochemistry, 200X). (b) Tumor cells positive for S100 (DAB, Immunohistochemistry, 400X). (c) Tumor cells positive for CD68 (DAB, immunohistochemistry, 200X). (d) Tumor cells negative for Langerin (DAB, Immunohistochemistry, 100X)

Papules as well as from the right axillary ulcerative lesion revealed intermediate-sized cells arranged in nodules in the dermis and in perivascular locations. These cells had bland chromatin with nuclear indentation and grooves with abundant cytoplasm. Epidermotropism was present with occasional mitosis. No eosinophils were seen [Figure 2a and b]. On immunohistochemistry (IHC), the atypical cells were strongly positive for CD1a [Figure 3a]. S100 and CD4 were strongly positive in a few of these cells and, CD68 was positive in few superficial cells only [Figure 3b and c]. Langerin and myeloperoxidase (MPO) were negative in these cells [Figure 3d].

Electron microscopy was not done as the patient refused a repeat biopsy.

A final diagnosis of IDCT was made based on clinical, histopathological, and immunohistochemical findings.

The patient refused treatment but his condition was nonprogressive even after 1 year of presentation.

Discussion

Indeterminate cell histiocytosis was first described by Wood et al. in 1985 as a neoplastic disease originating from dermal indeterminate cells that are characteristically positive for S-100 and CD1a but lack Birbeck granules. Histologically, IDCT is characterized by a dermal infiltrate composed of cells with abundant eosinophilic cytoplasm and oval-to-indented nuclei that resemble Langerhans cells. Immunohistochemical staining makes the distinction from other types of histiocytosis possible.[3]

Osseous involvement in an infant and corneal involvement have been reported.[4] Cutaneous involvement with pulmonary emphysema has been reported in an elderly lady.[5] Although viscera such as the kidney, liver, lung and, spleen can be involved in IDCT, there are no case reports so far of such involvement even after doing a thorough literature search.[6]

Uniform expression of CD1a and S100 protein enables the distinction of IDCT from other forms of non-Langerhans cell neoplasms. Differential diagnosis also includes Langerhans cell lineage tumors, such as Langerhans cell histiocytosis (LCH) and Langerhans cell sarcoma (LCS). LCH is a rare condition and mostly occurs in childhood. Unlike IDCT, LCH shows evident epidermotropism with intraepidermal Langerhans cell microabscesses. The presence of eosinophilic infiltration is another important difference from IDCT. However, immunpositivity for S100 protein, CD1a, and Langerin (CD207), and the presence of Birbeck granules confirm the diagnosis of LCH.[3] In contrast to Langerhans cell histiocytosis, IDCT is limited to the skin in the majority of cases (88%) and generally follows an indolent clinical course. It is important to differentiate it from LCH particularly to avoid aggressive overtreatment.[7] LCS, another important tumor in the differential diagnosis, is a very rare high-grade neoplasm where, unlike in IDCT, epidermotropism and eosinophil infiltration are not usually evident.[3] Myeloid sarcoma, which is an extramedullary tumor mass of neoplastic immature myeloid cells had been ruled out based on negative blasts in the peripheral blood smear and negative expression of MPO in the atypical cells.
In our case, even though eosinophils were not seen, epidermotropism was seen and the immunopositivity of CD1a and S100 with negative Langerin expression was crucial for definite diagnosis of IDCT.

**Conclusion**

IDCT with multisystemic involvement has been rarely reported. We, herein, report a case of IDCT with the possibility of multiorgan involvement. No direct mortality due to IDCT has been reported so far. Our patient, with extensive skin, liver, and lung involvement survived and the condition was nonprogressive even after 1 year of onset of the lesions without taking treatment.

**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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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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