

Primary Sino-Nasal T Cell Lymphoma in a Young Female: A Rare Case Report

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ABSTRACT

Sino-nasal lymphoma is a rare tumor of the elderly, rare in females and extremely rarely seen in young patients. It usually appears as a locally destructive mass with non specific symptoms and diagnosed by histopathology and immunohistochemistry. The tumor is treated with a combination of radiotherapy and chemotherapy and has a poor prognosis. We present an extremely rare case of T cell NHL in a girl of 16 years age. Present case highlights the importance of clinical suspicion in sino-nasal NHLs of young patients because early management can significantly improve the prognosis and survival of the patient.

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Introduction

Malignant lymphomas can arise in any part of the body and they are classified into hodgkins and non-hodgkins type. Primary sino-nasal lymphoma is a rare entity and usually of non hodgkins type. 10% of all primary non hodgkins lymphomas (NHL) arise in the head and neck.^[1] Among the subtypes seen, natural killer (NK)/T cell NHLs are more common in Asian population and B cell lymphomas are more common in western population. The tumor usually affects the adults and elderly with a male predominance but rarely can affect the childrens and young adults. Sinonasal lymphomas have a worst prognosis compared to other lymphomas occurring in other part of the body. Early diagnosis of the tumor is important because large size of the tumor is directly related to its poorer prognosis. We present a rare case of a young girl with primary sinonasal T cell NHL of the maxillary sinus and briefly discuss the management.

Case Report

A sixteen years old girl came to our otorhinolaryngology clinic with complaints of unilateral nasal obstruction, pain and puffiness in half of her face.(Fig 1a) Anterior rhinoscopy revealed a mass present in the right nasal cavity, which was whitish in color and firm in consistency. The patient didn't have any lymphadenopathy or any other mass present in her body.

CT scan showed a large heterogenous enhancing mass present in the right maxillary sinus, extending to infratemporal fossa, involving massater muscle with loss of interface of pterygoid muscle, extending into apex of orbit, retro-orbital fossa with loss of interface of rectus muscle, optic nerve and with erosion and destruction of posterolateral, anterior and medial wall of maxillary sinus. (Fig 1b & c) The mass was also involving maxillary antrum, right nasal cavity, nasopharynx and pterigopalatine fossa.

FNAC was done from the mass palpable through cheek and the smears showed heterogenous population of lymphoid cells comprising of small lymphocytes and some atypical looking lymphoid cells, some histiocytes and some neutrophils. The atypical lymphoid cells had enlarged, hyperchromatic and convoluted nuclei with scanty cytoplasm.(Fig 1d) A possibility of atypical lymphoproliferative disorder was suggested on the basis of cytological findings and the patient was advised an early biopsy. Histopathological section of the biopsy specimen showed diffuse proliferation of small to medium lymphoid cells having convoluted nuclei and coarse nuclear chromatin.(Fig 2a) The tumor was positive for CD 45 and CD 3 but was negative for CD 56 and CD 20. (Fig 2b, 2c & 2d) Depending upon the histopathological and

immunohistochemical findings the tumor was diagnosed as T cell NHL of maxillary sinus which was extending into the nasal cavity. No tumor extension was noted on peripheral blood examination, bone marrow examination and radiological examination of chest and abdomen.

The patient received a combination of radiotherapy and chemotherapy. She responded well and her tumor was reduced in size without any evidence of distant metastasis after six months of follow up.

Discussion

Primary malignant lymphomas of nose and paranasal sinuses are rare and accounts for 8% of all paranasal malignancies and 2% of all extranodal NHLs.^[1] Extranodal lymphomas predominantly arise in the liver, soft tissue, dura and bone marrow.^[1] Nasal NK/T cell lymphoma is a locally destructive tumor and was previously called as lethal midline granuloma, polymorphic reticulosis, midline malignant reticulosis, idiopathic midline destructive disease and lymphomatoid granulomatosis.

The common presenting symptoms of sino-nasal NHLs are nasal obstruction, nasal discharge, epistaxis, unilateral cheek swelling and headache. Though not in our case, non-specific symptoms are commonly observed in NK/T cell lymphomas. These include - fever, weight loss, weakness, night sweat, increased susceptibility to infection, peripheral lymphadenopathy and splenomegaly. These non-specific symptoms are due to cytokine secretion by tumor cells and more commonly observed in NK/T cell lymphomas.^[2]

Early diagnosis of the tumor is difficult because the tumor develops in an anatomical space. The tumor remains undetectable until it becomes progressive and starts involving adjacent structures^[1] as also seen in in our case. The tumor is located in the submucosa. It is non ulcerative on inspection and easily differentiated from squamous cell carcinoma but smooth surface makes it difficult to differentiate from minor salivary gland tumors and soft tissue sarcomas.^[2,3] Possible etiological associations are immune deficiency, Epstein-Barr virus infection and infection by Human T lymphotropic virus.^[4,5,6]

CT scan of the tumor shows mild to moderate enhancement in the involved soft tissue. C. H Ou *et al.* conducted a study on CT and MRI findings of nasal NK/T cell lymphomas and documented that MRI is better than CT scan in demonstration of tumor lesion.^[7] Cytological smears of the tumor shows a monomorphic population of atypical lymphoid cells and histopathology of the tumor shows a diffuse proliferation of small sized atypical lymphoid cells. The tumor cells have rounded hyperchromatic nuclei and

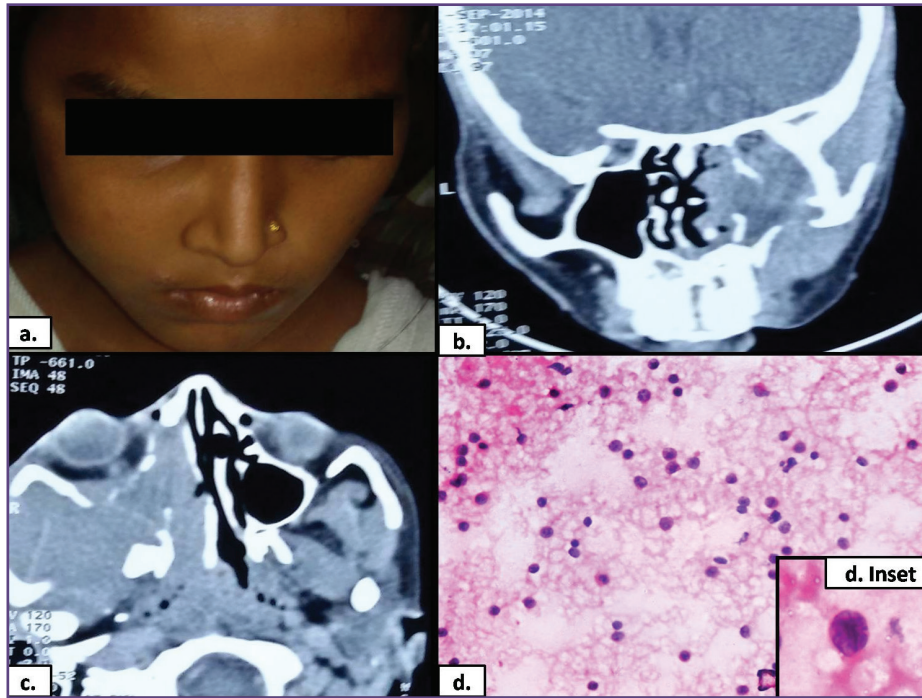


Fig. 1: (a) Clinical photograph of the case showing puffiness in half of her face. (b & c) CT scan showing a large heterogenous enhancing mass present in the right maxillary sinus, extending into apex of orbit, retro-orbital fossa, right nasal cavity and nasopharynx. (d.) FNAC smear of the mass showing atypical lymphoid cells having enlarged, hyperchromatic and convoluted nuclei with scanty cytoplasm (H & E 400X) (d. Inset) : H & E 1000X).

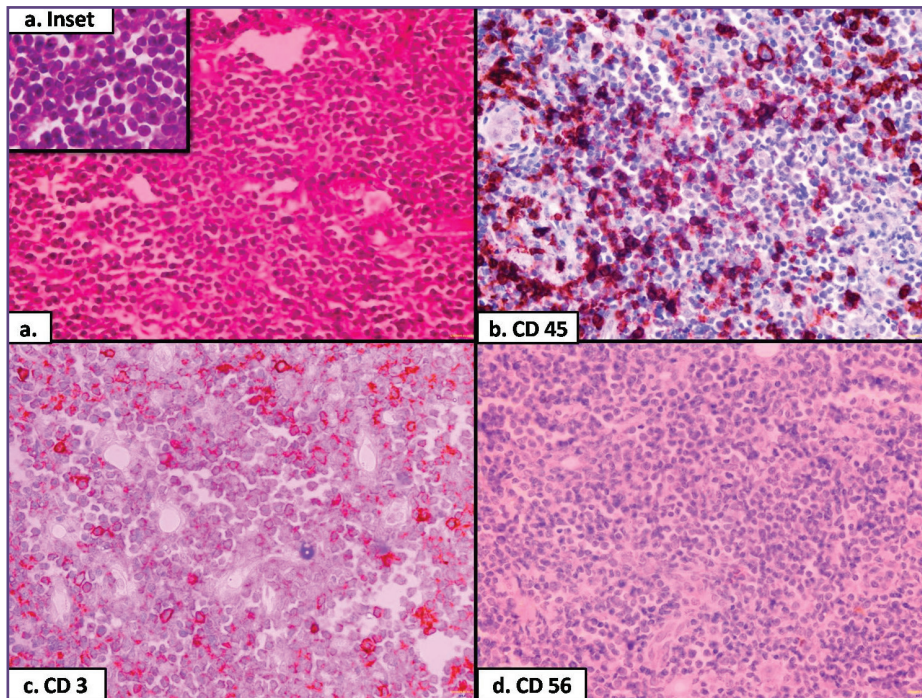


Fig. 2: (a) Histopathological section showing diffuse proliferation of small to medium lymphoid cells having convoluted nuclei and coarse nuclear chromatin. (H & E 100X) (a. Inset : H & E 400X). (b) CD 45 positivity by tumor cells. (c) CD 3 positivity by tumor cells. (d) CD 56 negativity by tumor cells.

coarse nuclear chromatin. The tumor shows positivity for CD 45 and CD 3 along with negativity for CD 20, indicating T cell origin. Sinonasal NK/T cell lymphomas show positivity for CD56 along with other T cell markers. Our case also showed positivity for CD 45 and CD 3, confirming the origin as T cell NHL. The microscopic differential diagnosis of sino-nasal T cell lymphomas are myeloid sarcoma, plasmacytoma, undifferentiated carcinoma and amelanotic melanoma.^[1] Microscopically, myeloid sarcoma is composed of medium to large hematopoietic cells with fine nuclear chromatin and prominent nucleoli; whereas plasmacytoma is composed of plasmacytoid cells. Undifferentiated carcinoma consists of uniform tumour cells with ovoid vesicular nuclei and prominent nucleoli along with an inflammatory component comprising of predominantly lymphocytes; whereas amelanotic melanoma is composed of epithelioid or spindle cells. On histological sections, plasmacytoid cells or any epithelioid or spindle cell morphology was not seen in our case and thus the possibility of plasmacytoma and amelanotic melanoma was ruled out; however the possibility of myeloid sarcoma and undifferentiated carcinoma could not be ruled out on histopathology. Immunohistochemically all of the tumours show negativity for CD45 and our case was easily differentiated from these tumours by immunohistochemistry as our case showed positivity for CD 45.

After diagnosis is made, a thorough physical examination, blood examination, radiological examination of chest and abdomen and bone marrow examination should be done to determine the tumor invasion.^[2]

The treatment of choice of large sino-nasal NHL is a combination of radiotherapy and chemotherapy. The most common chemotherapeutic regimen is cyclophosphamide, doxorubicin, vincristin and prednisone (CHOP) regime.^[6,8] Long term survival rate of the tumor is poor but a combined treatment of radiotherapy and chemotherapy has a better prognosis.^[3]

Conclusion

To conclude, primary sino-nasal T cell NHL is a rare tumor of elderly male and its occurrence in a young female is extremely unusual. The tumor shows a poorer prognosis

and its early diagnosis is important for better outcome and long term survival of the patients.

Funding

None

Competing Interests

None Declared

Reference

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