A Rare Case of Nasal NK/T-Cell Lymphoma in a Kazakh Man

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A 23 year old Kazakh man presented to the Republic Somatic Hospital in Semey, Kazakhstan, on 28 December 2017 because of progressive, painful destruction of the right half of his nose wing and septum. He experienced necrotic purulent nasal discharge with fetid odor, pain in the right maxillary sinus and infraorbital areas, and persistent headaches. His medical history revealed a diagnosis of chronic sinusitis 10 years before, which was treated with antibiotics, and a dull nose trauma 1 year earlier. Four months before presenting to the hospital, the patient had noticed swelling on the right side of the face, and people in close contact with him had begun to smell a fetid odor from his nose. Three weeks before presentation, a defect of approximately 0.5 cm in diameter had appeared on the right wing region of his nose, which rapidly expanded and spread to the nasal septum.

Physical examination showed complete absence of the right half of the nose, with a perforating, granulated wound extending to the soft tissues of the right cheek and reaching the midline of the nose and nasal septum. The medial wall of the right maxillary sinus, the nose septum, the bones of the latticed labyrinth, and the nasal conchae were all involved by the process.

Laboratory investigations did not reveal any abnormalities, and culture of nasal discharge was negative. Syphilis, granulomatosis with polyangiitis, and tuberculosis were excluded by laboratory and radiological exams.

A computed tomography (CT) scan conducted in February 2018 demonstrated destruction of the medial wall of the right maxillary sinus.

Histological examination of a biopsy of the nasal mucosa conducted in March 2018 showed individual glands incompletely covered with multilayer flat epithelium that was partly necrotic, stroma with abundant diffuse infiltration of lymphocytes, plasma cells, histiocytes, and leukocytes with the presence of a few large atypical cells with hyperchromatic nuclei. In the stroma, a large number of vessels with swollen endothelium and leukocyte infiltration of the walls was present [Figure 1A]. Immunohistochemistry showed expression of CD2 [Figure 1B], CD3, CD5, CD56. A diagnosis of nasal natural killer (NK)/T-cell lymphoma was made.

The lesions rapidly progressed and in May 2018 a CT scan revealed massive destruction of the medial wall of the right maxillary sinus, nasal bones, nasal septum, and right wing of the nose with destruction of the soft tissues of the nasolabial triangle and of the nose on the right [Figure 2]. The patient was treated with tramadol and prednisolone (70 mg daily) and radiotherapy was started soon after.

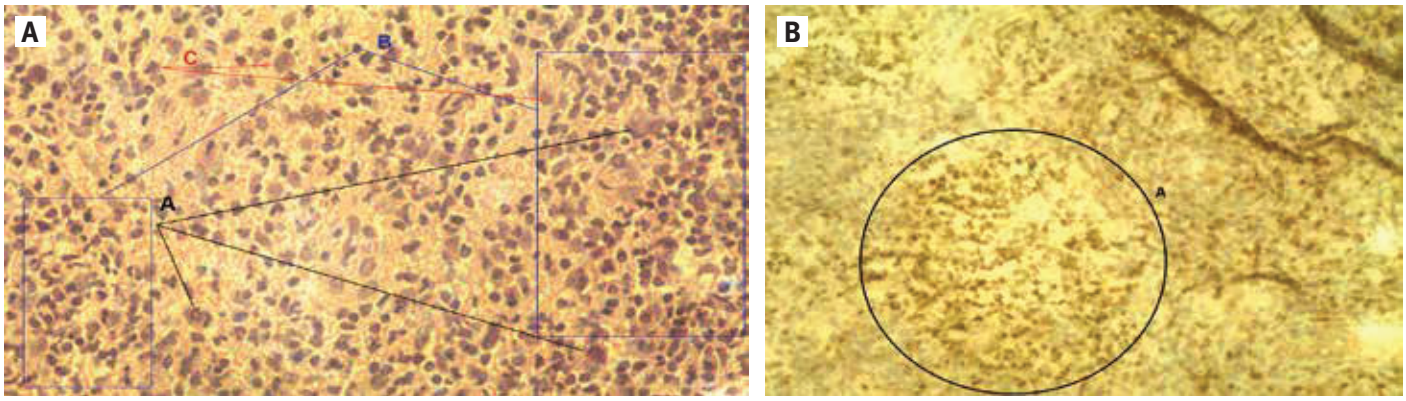
Primary lymphomas of the nose are rare cancers and were clearly described for the first time in 1966 [1]. They occur more frequently in Southeast Asia, Mexico, Latin America, and Indonesia than in Western countries [2]. They are more common in males, especially those in their fourth and fifth decades of life. The most common types are the extranodal NK/T cell lymphomas [3], which were once called lethal midline granuloma, polymorphic reticulosis, and malignant midline reticulosis.

The disease is characterized by necrosis that begins in the nasal cavity and extends centrifugally to mid-facial bones. The course is aggressive and often lethal, with destruction of the nose and face (midline) and a 5 year overall survival rate between 10% and 45% [4]. Death is often caused by septicemia, perforation into blood vessels, or penetration into the brain with ensuing abscess.

The pathogenesis is unknown, although there is an association with Epstein-Barr virus infection [5]. The histological pattern is characterized by a polymorphous proliferation of large, atypical, mono- or multinucleated cells, as well as small lymphocytes, plasma cells, neutrophils, and macrophages. Less frequently, eosinophils are present.

Therapy is rarely effective, with high relapse rates. Hematologists, oncologists, and radiation oncologists need to be involved in treatment. A standard treatment is still lacking due to the rarity of the

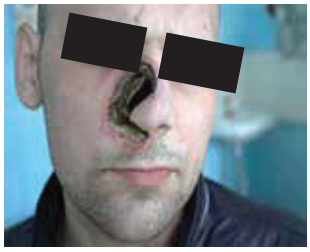
Figure 1. Histology of the nasal mucosa



[A] Black lines (A) point to atypical histiocytes with abnormal mitoses. Blue areas (B) show polymorphonuclear and immature lymphoid cell infiltrate. Red lines (C) lines point to atypical plasma cells (hematoxylin and eosin stain, 10×)

[B] Polymorphonuclear and immature lymphoid cells infiltrate. Immunohistochemistry shows CD2-expressing tumor cells (particularly concentrated within A circle) (100×)

Figure 2. Picture of the patient in May 2018



condition. Surgery may induce rapid progression of the disease [4]. A combination chemotherapy (e.g., dexamethasone, etoposide, ifosfamide, carboplatin) followed by external beam radiotherapy may be effective in patients younger than 60 years of age. Radiotherapy alone may be used in less advanced stages with a dose of 52 Gray.

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Capsule

Are diffuse and limited juvenile systemic sclerosis different in clinical presentation? Clinical characteristics of a juvenile systemic sclerosis cohort

Juvenile systemic sclerosis is an orphan disease. Currently, the majority of juvenile systemic sclerosis cohort studies are retrospective in design without standardized assessment. **Foeldvari** prospectively investigated the difference in manifestations of limited cutaneous juvenile systemic sclerosis and diffuse cutaneous juvenile systemic sclerosis subtypes. An additional aim was to compare these data to other juvenile systemic sclerosis cohorts and a large adult systemic sclerosis cohort. Patients fulfilling the Paediatric Rheumatology European Society juvenile systemic sclerosis classification criteria were included. Clinical characteristics and patient-related outcomes were assessed. In all, 88 patients with a mean disease duration of 3.5 years were enrolled: 72.5% with diffuse cutaneous juvenile systemic sclerosis with a mean modified Rodnan Skin score of 18 and 27.5% with limited cutaneous juvenile systemic sclerosis with mean modified Rodnan Skin score of 9. The mean age at the onset of Raynaud's and first non-Raynaud's symptoms was similar in both groups, approximately 9 and 10.5 years. Active digital tip ulcerations were present in 29% diffuse cutaneous juvenile systemic sclerosis and none in the limited cutaneous juvenile systemic sclerosis subjects (P = 0.005). Of those with cardiopulmonary testing, 3% of diffuse cutaneous juvenile systemic sclerosis and 23% of limited cutaneous juvenile systemic sclerosis group had cardiac involvement (P = 0.015), and 41% diffuse cutaneous juvenile systemic sclerosis and 22% of the limited cutaneous juvenile systemic sclerosis group had pulmonary involvement (P = 0.009). Physician global disease damage assessment was higher in the diffuse cutaneous juvenile systemic sclerosis group compared to the limited cutaneous juvenile systemic sclerosis group: 35 and 15 (P = 0.021).

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