CASE REPORT

Primary extranodal Natural Killer/T-cell lymphoma of the ethmoid sinus masquerading as orbital cellulitis

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ABSTRACT

This report presents a case of an exceptionally rare primary Natural Killer/T cell (NK/T) lymphoma of the right paranasal frontal and ethmoid sinuses in a patient treated previously for right side chronic sinusitis. It highlighted the importance of adequate tissue biopsy and pathohistological examination in patients with chronic sinusitis or orbital cellulitis that fail to respond to traditional management.

Key words: NK/T cell lymphoma, paranasal sinuses

INTRODUCTION

In 1897 McBride first described a patient with surface crusting, widespread necrosis and inflammation, aggressive and rapid destruction of nose and face midline, and lethal course (1). In the past, the term “lethal midline granuloma” was usually used but it included three histologically different lesions: Wegener’s granulomatosis, polymorphic reticulosis and malignant lymphoma (2).

Neoplasms of paranasal sinuses are very rare, comprising less than 3% of all autodigestive tract tumors (3). Lymphoma of the primary paranasal sinuses are even rarer and represent only 0-17% of all lymphomas in Kiel Lymph Node registry and account for only 5-8% of the extranodal lymphomas of the head and neck area (3).

Based on morphology and cell lineage there are currently 3 types of lymphoma: B cell, T cell and Hodgkin lymphoma. In addition, many proliferating T cells have shown to express an additional marker (CD56), which suggests an NK cell origin. These tumors are classified as NK/T lymphomas (4).

Most common presenting signs and symptoms of primary paranasal sinuses lymphoma are nonspecific and fall into several categories: nasal: epistaxis, nasal obstructions, congestion, extension into the nasal cavity; facial: unilateral facial or cheek swelling, facial asymmetry, pain, infraorbital nerve hypoesthesia; and ocular: unilateral tearing, diplopia, fullness of lids, pain, and exophthalmia (5).

In this paper we presented an uncommon case of primary NK/T lymphoma of the ethmoid sinus masquerading as chronic rhinosinusitis and orbital cellulitis.

CASE REPORT

A 60 year-old man presented with progressive well marked periorbital edema and erythema of the right eye, and thickness of the nasal dorsum and right cantal region. During the last 5 years he had been treated for right side chronic sinusitis. He had six millimeters proptosis and relative upper eyelid ptosis of the right eye. Nasal endoscopy revealed anterior deviation of nasal septum, obstructed ostiomeatal complex with a black mass and purulent discharge in the middle and common meatus.

Computed tomography (CT) scan of the paranasal sinuses and orbit showed soft tissue mass filling the right ethmoid, frontal and maxillary sinus, eroding the anterior part of the right lamina papiracea, and infiltrating right medial rectus muscle (Figure 1). Retention cyst in max-
illary sinus, polypoid mucosa of ethmoid sinuses and orbital soft tissue swelling without focal abscess were found during a functional endoscopic surgery. All of necrotic tissue was removed and first histological examination showed chronic inflammation of paranasal sinus mucosa. Bacterial and fungal cultures were negative. Four months later the patient developed fever, swelling, surface crusting, and widespread necrosis of the right periorbital and nasal area (Figure 2). Multiple biopsies of the paranasal sinuses were performed and diagnosed as nonspecific granulomatous inflammation.

Finally, diagnosis of NK/T cell (CD 56+) lymphoma was made by histological and immunohistochemical reexamination of the paraffine-embedded tissue obtained from the first biopsy of the ethmoid sinus and orbit. There were necrotic changes of varying degrees and a polymorphous pattern of proliferation involving large atypical cells with an occasional multilobated nucleus and various numbers of lymphocytes, plasma cells and macrophages. Features of vascular invasion by neoplastic lymphocytes were apparent. Occasionally, angiocentric pattern of proliferation was observed. Large atypical cells were positive for the NK-cell marker CD 56 (Figure 3). Patient had IV-A stage lymphoma and was EBV positive. Neck lymph nodes were negative. Thoracic and abdominal CT scans as well as bone marrow biopsy were all negative. He was scheduled for continuous chemotherapy with 8 CHOP 14-day cycles. Unfortunately, the patient’s condition deteriorated rapidly after the development of liver failure and respiratory failure and after 18 months he died.

Primary paranasal sinus NK/T-cell (CD 56 positive) lymphoma is a polymorphous extranodal lymphoma, expressing NK or rarely cytotoxic T-cell phenotype (3). It is an uncommon disease, and generally highly aggressive in its clinical course. Primary paranasal sinus T-cell lymphomas are much more frequent in Asian and Latin American countries, present at younger age, and usually arise from nasal cavity than the paranasal sinuses. In contrast, lymphomas of B-cell phenotype predominate in Western population and usually arise from paranasal sinuses. T-cell lymphomas are characterized by progressive ulceration and necrosis that are not typical for B-cell lymphomas (3, 5). Numerous studies showed that patients with NK/T lymphomas of the sinusonasal area had a high incidence of Epstein-Barr virus infections (6).

The majority of lymphomas involving the ocular adnexa are of B-cell lineage. However, NK/T-cell lymphoma is associated only infrequently with orbital or adnexal involvement (7). Life style and environmental factors significantly increased risk for developing NK/T-cell lymphoma among individuals exposed to pesticides (8).

Figure 1. Axial CT scan showing soft tissue mass in the right ethmoid, frontal, and maxillary sinus eroding bony structures and infiltrating right medial rectus muscle (D. Đanić, 2007.)

Figure 2. Widespread necrosis of the right periorbital and nasal area in a patient with primary paranasal NK/T-cell lymphoma (D. Đanić, 2007., with patient’s permission)
This patient lived in a rural area, worked in agricultural environment and was exposed to pesticides for many years.

The diagnosis of lymphoma cannot be made from clinical findings solely and thus biopsy and imaging of the lesions is mandatory prior to any treatment (9). During the diagnostic procedure adequate tissue biopsy must be taken to differentiate lymphoma from destructive inflammatory diseases or malignant tumors (9). Biopsy must be adequate, not too small or too superficial because sinusonasal lymphomas are subepithelial lesions, often with perfectly normal overlying mucosa, unlike carcinoma, which are usually ulcerative (9). Repeated biopsy may sometimes be needed. Cross-sectional imaging findings like pathologic contrast enhancement or bone changes may reveal the malignant nature of the disease but there is significant overlapping between those possible pathologies that can arise in this region (9). Physiologic imaging, like perfusion CT and proton MR spectroscopy, in the extracranial head and neck can be implemented in any CT or MRI survey, provide functional information of the lesion, and may be helpful to differentiate benign from malignant disease as well as guide therapeutic decisions (10).

The optimal treatment for primary nasal lymphoma remains unknown (11). Surgical resection of paranasal sinusal lymphoma is not recommended unless the tumor spreads to critical locations resulting in impending death (11). Complete response rate after radiotherapy is much higher as compared to chemotherapy although radiotherapy planning for primary nasal lymphomas may be difficult because these lymphomas often encroach on such radiosensitive critical structures as the optic chiasm, optic nerve and eyeballs and exact dose-tumor response relationship is unknown (11). Addition of chemotherapy to radiotherapy did not improve survival rate with early stage NK/T cell lymphoma (12). Ocular manifestation prior to systemic ones may be useful to monitor the response to therapy (12).

Prognosis associated with sinonasal NK/T cell lymphomas varies. Dissemination is infrequent, but when it occurs it typically involves other extranodal sites (12).

In conclusion, this case highlights the importance of adequate tissue biopsy and pathohistological examination in patients with chronic sinusitis or orbital cellulitis that fail to respond to traditional management.

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REFERENCES

Knee disarticulation

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ABSTRACT

In this paper we presented three patients with knee disarticulation performed according to Baumgartner. The Baumgartner technique and the application of knee disarticulation prosthesis appeared to be superior in comparison with other methods.

Key words: knee, disarticulation, Baumgartner technique

INTRODUCTION

Knee disarticulation is a rarely used method in amputation surgery, primarily due to the operative technique itself and secondly, due to poor understanding of prosthetic replacement possibilities (1). Technological progress and new developments in the prosthetics have opened new possibilities of amputation methods and consequently in the choice of the amputation level (1). A prosthesis for patients with knee disarticulation has been designed, with construction being based on the operative method of knee disarticulation according to Baumgartner (2). The energy expenditure during walking with knee disarticulation prosthesis is a little more than 40%, the same as for below-knee prosthesis (3).

Disarticulation of a knee is recommended for high traumatic amputation of the below-knee, crush injury, complex injuries and tumors of the below-knee (1). Surgeons have been in dilemma between the method of transcondylar amputation and knee disarticulation (2). Knee disarticulation proves to be superior due to the possibilities of prosthetic replacement. The advantages are: a long and strong stump with a tip that can endure full-weight bearing and is suitable for a knee disarticulation prosthesis, the energy expenditure during walking equal to walking with below-knee prosthesis, normal function of the above-knee muscles (2). Unpopularity of knee amputation over many years was caused by bad experience with primary wound healing and the resulting stump of poor quality with regard to its function (2). In order to prevent these complications some surgeons introduced modifications in operative method (4-6). These methods are surgically more demanding and associated with a higher risk of complications (7).

Baumgartner 1971 describes the method of knee disarticulation as a surgically simple procedure that creates a functionally satisfactory stump with regard to further prosthetic fitting (2). The simplicity of the technique is reflected in every aspect - skin, cartilage, bone, muscles (2).

During the last 10 years the Clinic of Traumatology Zagreb has been using the technique of knee disarticulation described by Baumgartner. However, we have introduced some minor modifications. Instead of sutturing the patellar ligament as Baumgartner was practising, we cut ligament at the top of the patella. So we additionally increased the contact and weight-bearing surface...