Primary B cell lymphoma of paranasal sinuses: a diagnostic surprise

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Abstract Primary lymphomas of paranasal cavities are rather uncommon entities. They have a variable presentation from fulminant destructive manifestations to chronic indolent type of disease. Chronic indolent form may mimic invasive fungal sinusitis in its presentation. Unless high index of suspicion is held and appropriate histopathology sections are taken from specimen, its diagnosis can be deceitful. We here by report a case of primary lymphoma of the paranasal sinuses which was radiologically and clinically suspected to be a invasive fungal sinusitis and later was proven to be a B cell lymphoma. Clinical similarities between lymphoma and invasive fungal sinusitis along with management issues are discussed in this article.

Keywords Paranasal cavities · B cell lymphoma · Non-hodgkins Lymphoma (NHL)

Introduction

Non-hodgkins Lymphoma (NHL) of the sinonasal tract is rare in the western world and accounts for only 0.17% of all lymphomas in Kiel Lymph Node Registry [1]. The majority of cases are B cell type [2]. In Asia, sinonasal lymphoma is more common, accounting for 7% of all NHLs, and it is more likely to be of the Natural Killer(NK)/T-cell variety rather than B-cell [2].

NHLs are a heterogeneous group of diseases arising in nodal or extranodal lymphoid tissue. A favored presentation site is the head and neck, where between one-third and two-thirds of all extranodal NHLs arise. The spectrum of biologic behavior in head and neck lymphomas ranges from indolent tumors, present for years before diagnosis, to rapidly growing tumors that double in size over days. The distinction in cell type is important because of significant differences in natural history and response to treatment. Both B-cell and NK/T-cell lymphomas in this location are aggressive, but NK/T-cell tumors are more likely to arise in the nasal cavity, affect younger patients, and have an aggressive local course with an extremely poor prognosis. An early diagnosis of primary NHL of the sinonasal tract is unusual because such a lesion develops in an anatomic space and expands toward the sinus or nasal cavity, usually causing no symptoms at the early stages [4]. Only after reaching a considerable size and involving adjacent anatomic structures do the presenting symptoms appear, and they may masquerade as other nasal or head and neck findings. Presenting symptoms depend on the site of origin within the sinonasal tract and include nasal passage obstruction, epistaxis, ulceration, cranial nerve deficits, facial swelling, proptosis, and pain [5]. Radiological impression of the lymphoma in CT scan of paranasal sinus may often mimic invasive fungal sinusitis. Bone erosion of paranasal sinuses and orbit hence should be seen with great caution, as they can be signs of malignancy.

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Case report

A 30-year-female presented with right nasal obstruction for six months and headache for three months. There was history of purulent posterior nasal discharge but there was no history of epistaxis. She also complained of fullness of ear on right side. On clinical examination right side nasal cavity revealed a polypoidal mass which was dirty white in colour. It was firm on probing and did not bleed on touch.

Fig. 1 Plain CT scan in coronal section shows heterogeneous mass occupying left maxillary sinus, anterior and posterior ethmoid sinus and sphenoid sinus. Sphenoid septum is eroded and it is extending in the right side sphenoid sinus. Right side maxillary sinus is also involved

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Bilateral ears had otitis media with effusion. Eye and its appendages were normal. No proptosis or restricted eye movements were noted. There was no lymphadenopathy. Nasal endoscopy revealed a polypoidal mass arising from right ethmoids and middle turbinate. It was encroaching the choana and nasopharynx. Base of skull was free. A provisional diagnosis of fungal sinusitis was made. CTscan revealed a heterogeneous mass arising from paranasal sinuses involving right side maxillary, ethmoid, sphenoid sinus and nasopharynx. There was minimal bone erosion of sphenoid floor and posterior ethmoids. However orbits and lamina papyracea were normal. Patient underwent Endoscopic sinus surgery. Intraoperatively dirty white polypoidal mass along with whitish cheesy material was found filling the right maxillary, ethmoid and sphenoid sinuses. The appearance was different from that of allergic mucin. The entire specimen was collected and sent for histopathology and fungal culture. The pathologic examination revealed it to be primary non-Hodgkin's lymphoma of the paranasal sinuses. The KOH staining and fungal culture did not reveal any hyphae. Immunohistochemistry staining showed positive markers for CD20, CD45 and confirmed it to be a primary B cell NHL of paranasal sinuses.

Patient underwent 6 cycles of chemotherapy with cyclophosphamide, adriamycin and vincristine and showed complete response. Patient is asymptomatic for last four months and post chemotherapy nasal endoscopy did not show any mass in the nasal cavity.

Discussion

Non-Hodgkin's lymphoma of the sinonasal tract is rare in the Western World, where the majority of cases are of the B-cell phenotype. Cases can occur over a wide range of ages, peaking in the sixth decade, with males overall predominating over females. B-cell lymphomas are more likely to have a primary site of origin in the paranasal sinuses and present as a mass causing obstructive symptoms. Typical presenting symptoms include nasal obstruction, nasal discharge, epistaxis, and pain or swelling of the nose or face. Patients may also demonstrate signs due to infiltration of local structures, such as cranial nerve palsies or diplopia and proptosis due to intra-orbital or skull base extension. In contrast, T cell and NK cell NHL are more likely to have a primary site of origin in the nasal cavity and to present as destructive lesions such as septal perforations, non-healing ulcers and fistulae [6]. The radiographic patterns are typical of bulky tumors with local infiltrative growth resulting in prominent bony erosion of the sinus walls. High-grade B-cell tumors have been noted to cause orbital soft tissue and osseous destruction, whereas nasal septal perforation or destruction is more frequently associated with high grade T-cell lesions. However in our case no such involvement was seen. This may be presumably because of early detection done in our case.

Radiologically, a definite mass, or, less often, cloudy or opaque sinuses are apparent. Computed tomographic scans are particularly helpful in evaluating the extent of the destruction and the involvement of the sinuses and orbits and should be considered the gold standard investigation [4]. Although T2-weighted magnetic resonance imaging (MRI) enables us to distinguish tumour from thickened mucosa or obstructed fluid [3], MRI is not a routine investigation for pre-operation evaluation of sinus surgery patients. In our case, CT scans of the sinuses showed soft tissue attenuation of ethmoid and sphenoid sinus and obstruction of the right osteomeatal complex suggestive of fungal sinusitis.

Considering the young age of the patient and the radiologic picture, the fungal sinusitis was kept as the probable diagnosis. It was only after the post operative histopathologic examination that the possibility of lymphoma came to light.

In our case we would like to emphasise the need for histopathological examination of all nasal masses. There is no general consensus about the need for routine histopathological examination of nasal polyps among otolaryngologists. Alun-Jones *et al.* described only 38 per cent of ENT surgeons practiced it routinely in the UK and suggested that it was unnecessary to send all nasal polyps for pathological examination. As in this case the clinical picture and radiologic investigation alone may lead us to deceptive diagnosis and thus tissue diagnosis becomes imperative to rule out such rare malignancies. Lumsden *et al.* reviewed 18 cases of unusual polypoidal tumours of the nasal cavity and suggested that unexpected neoplasms may present as polyps. Less recognized is the fact that so many of these nasal lesions may appear to be ‘simple polyps’ on anterior rhinoscopy but on examination turn out to be malignant on histopathology. Therefore it is important to submit all sample materials for histopathology.

Conclusion

This paper presents a case of non-Hodgkin’s lymphoma that was unexpectedly discovered during histopathological examination of tissue removed during surgery for rhinosinusitis. This finding reminds us that all polypoid tissue removed from the sinuses and nose during surgery ought to be submitted for histological examination.

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