Introduction

Fifty-five percent of lacrimal duct tumours are malignant, and most of these malignant tumours are epithelial tumours. Non-epithelial tumours account for just one-third of such malignant tumours. Plasmacytoma, which is a rare lesion and a type of malignant plasma cell tumour, has never been reported in the lacrimal duct. Plasma cell neoplasms have been classified as monoclonal gammopathies of undetermined significance, as plasma cell myeloma, as solitary plasmacytoma of bone, as extraosseous plasmacytoma (EOP), and as monoclonal immunoglobulin deposition disease. EOP is defined as a neoplastic proliferation of plasma cells arising outside the bone marrow and accounts for ~3-5% of all plasma cell tumours. EOPs are two times more common in men, and the mean age of affected patients is 55 years. Approximately 80% of EOPs are in the head and neck region and commonly affect the nasal cavity, the paranasal sinus, the tonsillar fossa, and the oral cavity. Patients typically present with localized disease that has an insidious natural clinical course. The etiology of this disease is still unknown, but chronic stimulation, overdose radiation, viral origin, and gene interactions in the reticuloendothelial system have been suggested as etiologic factors. Here we present a rare case of EOP in the lacrimal duct.

Case report

A 66-year-old man was referred to our institution for treatment of a right-sided nasal mass that was located between the internal canthus and the right nasal floor. The tumour had been growing slowly for a few months and did not cause the patient any pain. The patient specifically noted right nasal obstruction without any epistaxis or rhinorrhea. Apart from this, the patient had no relevant medical history. Clinical examination showed that the medialized lateral wall of the right nasal cavity completely obstructed the right nasal cavity. The mucosa lining appeared normal. CT scan and MRI were performed. CT scan showed a 5-centimeter large tissular mass involving the right lacrimal cyst and duct (Figure 1). The lesion was isointense on T1-weighted and hyperintense on T2-weighted MRIs (Figure 2). The lesion had a regular appearance with no sign of tumoural infiltration. These findings were consistent...
with the diagnosis of a slowly growing non-aggressive tumour.

Minimally invasive endoscopic nasal surgery was possible and was subsequently performed. The entire mass was removed, alleviating the nasal obstruction. Histological examination showed densely packed, round, polygonal cell structures scattered throughout relatively sparse stroma. We also observed marked proliferation of regular plasma cells without cytological abnormalities. These findings were consistent with the diagnosis of a solitary plasmacytoma. Immunohistochemical study showed tumour cells that were positive for CD45, CD138, and CD79a; there was slight positivity for Ki67. There was no monoclonal proliferation (heavy and light chains) as determined using immunofluorescence. The diagnosis was thus a solitary non-secreting plasmacytoma of the lacrimal duct.

Bone marrow aspiration and biopsy revealed the absence of plasma cell infiltration, and blood cell counts were normal. No monoclonal components were detected in plasma or urine after the surgery. Serum immunoglobulin kappa and lambda light chains levels were normal, with a k:l ratio of 1:1.

For complete staging, post-operative imaging examination by fusion positron emission tomography (PET) scanning was performed. PET images revealed a strong positive signal on the roof of the right maxillary sinus, suggesting a residual tumour in the proximal lacrimal duct, and a moderately positive signal on the right side of the neck, consistent with tumour activity in the lymph node. The lesion on the roof of the sinus was certainly a residual tumour that was still growing after the first surgery. However, the suspect cervical lymph node suggested metastatic spread, especially after a comparison of CT scans performed before the surgery and three months after the surgery showed that the suspect lymph nodes were not present at the initial presentation. At that time, i.e. three month after the first surgery, clinical examination did not show anything new: the nasal endoscopic examination revealed no lesions, and cervical palpation did not reveal any pathologic lymph nodes.

The multidisciplinary staff, which included hematologists and pathologists, made a joint decision regarding treatment, and surgery was performed to remove both the cervical lymphadenopathy and residual tumour in the roof of the sinus. It was important to
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make sure of the diagnosis of disseminated EOP without evolution towards multiple myeloma before initiating specific tumoural treatment. The histopathological findings were consistent with plasmacytoma localization both in the residual maxillary tumour and in the two cervical lymph nodes. Because of early spreading consistent with an aggressive case of EOP, the patient underwent 6 cycles of chemotherapy with bortezomid and dexamethasone followed by radiotherapy. Twenty-eight months later, the patient is still in remission with no signs of progression towards multiple myeloma.

Discussion

Solitary EOP is a localized plasma cell neoplasm. This plasma cell immunoproliferative monoclonal disease arises outside the bone marrow without clinical evidence of multiple myeloma. Specifically, EOP originates from a clone of malignant transformed plasma cells that migrate and settle in soft tissue or in an area where there is extracellular connective tissue. EOP accounts for fewer than 1% of all head and neck malignancies and for fewer than 4% of non-epithelial tumours of the nasal tract. EOP also accounts for 3-5% of all plasma cell neoplasms. The incidence is higher in men aged 60–80 years, with a male to female ratio of > 3:1. Most EOPs are located in the head and neck and involve the sinonasal or nasopharyngeal regions (75%), the larynx (18%), and the oropharynx (12%). Other rare localizations have been reported, including the tongue, tonsils, minor salivary glands, posterior pharyngeal wall, thyroid and parotid glands, and even the middle ear. To our knowledge, EOP in the lacrimal duct has not been reported previously.

Late diagnosis is common for EOP, since it presents as a slowly growing mass. In the case described here, the patient sought medical help only when the mass affected the right lateral nasal wall and progressively obstructed his right nasal passage. Epistaxis, nasal discharge, pain, cranial nerve palsy or cervical lymphadopathy are less common, as are non-specific findings in the initial presentation. Imaging using CT scanning and MRI aid in assessing local disease, but definitive diagnosis requires biopsy for histological and immunohistochemical investigation. A recent review reports that 55% of lacrimal cyst tumours are malignant, leading to death in 38%. Other reports do not show any predominance in terms of race or sex. In most cases, the initial diagnosis is incorrect; usually the tumour is wrongly diagnosed as dacryocystitis. Imaging modalities such as CT and MRI may allow clinicians to identify inflammation associated with the tumour, but imaging cannot determine the tumour type. Thus, early histological diagnosis from biopsy material remains the gold standard. Tumours in this location are often diagnosed in a later stage. In the case reported here, the mass was removed completely by endoscopic surgery to treat the nasal obstruction. Lacrimal cyst malignant tumours are mostly epithelial tumours (66%), including squamous cell carcinoma and adenocarcinoma. Non-epithelial tumours are mainly lymphomas, melanomas, sarcomas, or malignant granulomas. Plasmacytoma has not been described previously in reports of these non-epithelial malignant tumours of the lacrimal duct.

In its natural evolution, EOP can transform into multiple myeloma and has a conversion rate of 15-20%. This conversion is associated with a poorer prognosis. The conversion rate seems even more important if the EOP involves the adjacent bone. Local recurrence has been reported to be as high as 10%. Dissemination of the tumour can occur even after a long latency time, and this takes place in 35–50% of EOPs. In the present case, the patient experienced dissemination three months after the first surgery. This unusual and confusing clinical evolution led us to verify the tumour’s histologic features with the aid of several specialized pathologists. Despite this unusual clinical pattern, which mimicked a possible MALT lymphoma, pathologists noted that local recurrence and lymph node dissemination were in accordance with plasmacytoma proliferation. This case thus illustrates how clinical presentation can be misleading and emphasizes the need for information obtained through biopsy of the lesion.

For many clinicians, the treatment of choice for EOPs is radiotherapy, because this type of tumour is highly radiosensitive. However, there is no consensus regarding EOP treatment. Some reports describe long-term control following radiotherapy with a high likelihood of cure, and a recent study reports higher 5-year local recurrence-free rate in patients treated primarily with radiation. Surgical excision of the tumour does not appear to be common practice, even though some studies show that surgery can be
as effective as radiation therapy. Surgery also seems to be the treatment of choice for large tumors and for lesions with extensive bone destruction. For these reasons, surgery alone could have been a good choice for removing the initial lacrimal tumor, especially considering that alternative treatments such as radiotherapy can have side effects, and the lesion was situated close to the eyes of our patient. It seemed unlikely that the tumor was spreading. Surgery on the two additional lesions was necessary for histological assessment of dissemination. These observations highlighted the importance of performing a biopsy of the lesion and, depending on the staging results, of managing any suspect sinonasal lesion as cancer until proven otherwise.

Thus, the clinical features and histopathological findings in this patient were consistent with EOP stage II according to Megat Shiraz et al., which led us to propose chemotherapy and radiotherapy. Usually the prognosis for EOP is good, with a 5-year survival rate of 76%, but long-term follow-up is essential because local recurrence and disseminated disease can occur.

References


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