Preauricular malignant lymphoma masquerading as benign pleomorphic adenoma – a rare tricky presentation

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Abstract

Lymphoma in the head neck area most commonly present as cervical lymph node swelling followed by tonsillar enlargement but the presentation as sole preauricular swelling without systemic involvement is a very rare finding. Here we present a case of malignant lymphoma Grade I, presenting as a single preauricular swelling masquerading as a benign pleomorphic adenoma, in a 24 year old girl.

Keywords Malignant lymphoma · Pleomorphic adenoma · Preauricular region

Introduction

Lymphoma is the second most common tumor in the head neck area. They usually present as slow growing painless neck swelling followed by tonsillar swelling. Non-hodgkin lymphoma exceeds Hodgkin lymphoma in this area [1,2,3]. Primary preauricular presentation near the parotid area without any local or systemic lymphadenopathy masquerading as a benign pleomorphic adenoma is a very rare finding.

Case report

A 24-year-old girl presented to the out-patient of Department of Otolaryngology and Head Neck Surgery, Calcutta National Medical College with a complaint of swelling in the right preauricular region for the last one month. The swelling had an insidious onset, progressed fairly rapidly over the last one month. She gave no history of any systemic symptoms like fever, malaise, weakness or weight loss.

On examination of the patient, the swelling was seen to be reddish in colour, having a smooth surface (3 cm × 2 cm) with no evidence of overlying scar, sinus or fistula. On palpation, it was firm in consistency, mobile, adherent to the overlying skin and margins were distinct. It was nontender and having normal skin temperature. (Fig. 1)

On loco-regional examination, no palpable cervical lymph nodes were present and on cranial nerve examination no cranial nerve deficits were present.

The patient was admitted and subjected to routine investigations like blood tests, chest x-ray and some specific tests like FNAC. On FNAC, we got a diagnosis of pleomorphic adenoma. Depending on this report we decided to perform excision of the mass along with a random type flap repair to achieve good cosmetic outcome since it was present on the face of a young unmarried female.

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Fig. 1 Clinical photograph showing preauricular mass

Fig. 2 Photograph showing specimen of excised preauricular swelling

Fig. 3 Intra-operative photograph showing fashioning of rhomboid flap

Fig. 4 Photograph showing recurrence of swelling with flap necrosis

Fig. 5 Histopathological slide showing features of malignant lymphoma (H&E stain, 10X10)

Fig. 6 Histopathological slide showing features of malignant lymphoma (H&E stain, 10X40)

Operative Procedure

The patient was operated under general anesthesia. The head was turned to the opposite side and elevated to 15° to reduce venous congestion. The mass was removed completely by giving a circumscribed incision along with a complete tumor free margin. The mass had no relation to parotid gland (Fig. 2). The defect was covered with rhomboid shaped

random type of flap taken from below which was pulled anteriorly and forwards to cover the entire defect (Fig.3). Sutures were removed after two weeks. The specimen was sent for biopsy which again gave a diagnosis of hypercellular variety of pleomorphic adenoma thus confirming the previous FNAC report.

The patient came for follow up 2 weeks after discharge. On examination, the flap was showing early signs of necro-

sis like change of colour and unhealthy looking wound. The findings progressed over the subsequent week. A swelling which was identical but more extensive than the first one started to gradually appear around the excised area but now it was also involving the tragus (Fig. 4). On clinical suspicion, based on the flap necrosis, fixity and rapid recurrence we decided to review the slides which astoundingly gave a diagnosis of malignant non-hodgkin lymphoma small cell type which was contrary to the previous biopsy report (Fig. 5, 6). The paraffin blocks and slides were sent to another pathologist for thorough review who reconfirmed the diagnosis of malignant lymphoma. Thereafter we carried out USG, chest x-ray and CT scan of the abdomen to assess involvement of probable sites and extent. All tests were insignificant. So a diagnosis of malignant preauricular non-hodgkins lymphoma was arrived at and the patient was referred to the department of radiotherapy for subsequent management. Further follow-up for up to 6 months showed no obvious recurrence.

Discussion

Lymphomas are the second most common malignant tumor in the head neck area in adult patient preceded only by metastatic adenopathy. They present most commonly as a slowly progressing non-tender neck mass followed by intraoral swelling [1, 2, 3]. Primary presentation in the preauricular area without involvement of any local or systemic lymph nodes as present in our case is a rare finding. Incidence is generally more common in the males unlike in our case. They are usually slow-growing, firm, non-tender masses. According to established textbooks any lymph node in the neck larger than 10 mm qualifies as cervical lymphadenopathy. They most frequently start as enlarged lymph nodes in the neck, axillae or inguinal regions [1, 3].

There are two major kinds of lymphomas, hodgkin's and non-hodgkin's lymphomas. Hodgkin's has a bimodal peak and is usually associated with a good prognosis, while non-Hodgkin's lymphomas may occur at any age and are associated with a less favorable prognosis. The non-Hodgkin's lymphomas are a very heterogeneous group of diseases with a diverse pathology and natural history. They are divided into three major categories: the low grade, intermediate grade, and high-grade lymphomas [1, 4].

Till date there is no established cause for lymphomas. However, it is believed that they develop as the result of hereditary, genetic and environmental factors. Most patients have underlying genetic defects. Non-hodgkin's lymphoma appears to be more related to DNA mutations that are usually acquired after birth, rather than being inherited. Infection with the human T-cell leukemia/lymphoma virus (HTLV-1) increases a person's risk of developing certain types of T-cell, non-hodgkin's lymphoma. Lymphomas are staged according to the Ann Arbor staging which allows the treating physician to determine the precise strategy of treatment,

prognosis, and outcome of treatment. Histologically they are graded according to Rappaport classification [3, 4].

Fine needle aspiration biopsy gives an idea about the histology but an excision biopsy is the only way to reach an accurate definitive diagnosis [1, 3, 4, 5]. In our case on FNAC a diagnosis of pleomorphic adenoma was given depending on which we carried out the excision. This fallacy might be explained by the fact that the pathologist probably examined the underlying or associated salivary tissue and not the preauricular lymph nodes. This confusion may very well arise as they are in close contiguity and because lymphoma can in fact also arise from the intraparotid nodes.

After sending the excised specimen for biopsy we got a diagnosis of hypercellular variety of pleomorphic adenoma with excess mitosis but no frank malignant cells were seen. This histopathological report justified our excision but when the patient came at follow up with a very rapid recurrence and fast progression, we had an immediate clinical suspicion that something was wrong. Only then on reviewing the slides a diagnosis of malignant lymphoma was arrived at and the patient was referred to department of radiotherapy for further management. She is doing well with excellent response at 6 months follow up. After extensive research, we did come across few cases of such occurrence where hypercellular variety of pleomorphic adenoma caused confusion with lymphoma. But very interestingly none of them had a single preauricular presentation devoid of any other lymphadenopathy as seen in our case.

If histopathological examination of the biopsied tissue does not provide a definite answer, other laboratory tests are usually needed, such as immunohistochemistry, flow cytometry, cytogenetics, or molecular genetic studies. However, high costs and poor availability are major constraints. In addition USG, chest X-ray, CT scan, MRI or gallium and PET scan to help determine how far it may have spread and for staging [2, 3, 4].

These diseases are usually treated with chemotherapy and radiation. Surgery has a minimal role in treatment and its major role is confined to diagnosis and staging. Radiation therapy is frequently used for early Hodgkin's disease. In non-Hodgkin's lymphomas, the role of this approach to therapy is limited, and most patients with these diseases require systemic chemotherapy. There is a great deal of controversy as to the different ways of combining radiation and chemotherapy. However, Stage I of Hodgkin's disease may be treated by radiation therapy alone. This may also hold true for non-Hodgkin's lymphoma with Stage I and with no symptoms. In all other categories of diseases, systemic chemotherapy becomes indicated [4, 6].

In Hodgkin's lymphoma, two types most commonly used are MOPP and ABVD. The latter is preferred to the former because it is at least as effective but less toxic. In non-Hodgkin's lymphoma CHOP is the most commonly employed chemotherapeutic regimen. When patients fail to respond to CHOP, an alternative regimen will be ASHAP.

Usually 6–9 cycles of chemotherapy are needed to achieve a good control of the disease [4, 6].

Bone marrow transplantation is currently evaluated. Some recent developments such as monoclonal antibodies and radioimmunoglobulins have also shed new hopes of better control [6].

Conclusion

Malignant lymphoma though commonly presents as a painless slowly enlarging cervical mass or tonsillar swelling, it may indeed very rarely present as a very benign looking solitary preauricular swelling. Diagnosis of such a swelling can be very troublesome as it may cause histological confusion with hypercellular variety of pleomorphic adenoma. Even FNAC can be misleading and histopathological reporting also can be tricky. Hence every surgeon as well as pathologist should be very aware of such possibility, as mode of treatment is entirely different for lymphoma and pleomorphic adenoma

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