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Case Report

Carcinoma ex pleomorphic adenoma of the parotid gland: a case with difficult diagnosis

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ABSTRACT

Carcinoma ex pleomorphic adenoma (CXPA) is carcinoma arising from a primary or recurrent benign pleomorphic adenoma. It might be non-invasive or invasive. It often poses a diagnostic challenge to clinicians and pathologists. Pathological assessment is the gold standard for diagnosis. Treatment for CXPA often involves an ablative surgical procedure, which may be followed by radiotherapy. We report to you a 63-year-old female who came to the ENT OPD with a complaint of swelling in the front of the right ear for 6 months and pain over swelling for 2 weeks. The patient was advised for USG-guided FNAC, which was suggestive of a benign cystic parotid lesion. The patient was planned for superficial parotidectomy and the tumor was surgically excised. The frozen section was suggestive of a benign salivary gland tumor and the histopathology report concluded of a non-invasive carcinoma ex pleomorphic adenoma. The patient was advised to undergo radiotherapy after the surgery. During the follow-up, the patient showed signs of Frey's syndrome. Carcinoma ex pleomorphic adenoma is difficult to diagnose; clinicians and pathologists should work together to diagnose it and pathological assessment is the gold standard for the diagnosis.

Keywords: Carcinoma ex pleomorphic adenoma, CXPA, Non-invasive, Pleomorphic adenoma, Parotid gland

INTRODUCTION

Carcinoma ex pleomorphic adenoma (CXPA) is defined as a carcinoma arising from a primary or recurrent benign pleomorphic adenoma (PA).[1,2] Most patients present in the 6th and 7th decades. It shows a predilection toward females.[3] The classic case is a patient with a long-standing mass that suddenly undergoes rapid growth over a period of several months.[4] Parotid malignancies are rare and account for 1–3% of all head and neck cancers.[5] CXPA more commonly occurs in the major salivary glands than in the minor salivary glands. Further, in the major salivary glands, the parotid (67%) is the most frequently affected gland and the sublingual is the least affected (<1%).[6] The pathogenesis is uncertain, it could be a malignant transformation of a benign pleomorphic adenoma or could be a malignant entity from the start.[7] The malignant component should be classified as non-invasive, minimally invasive or invasive.[8] For those tumors that do not invade beyond the capsule or the rounded edge of the tumor, there is no risk of recurrence or metastasis from a completely resected tumor. For invasive tumors, those with high-grade features do poorly. Wide resection with lymph node dissection and radiotherapy (RT) is the treatment of choice.[9]

CASE REPORT

A 63-year-old female, presented with a six-month history of right parotid swelling and two weeks of associated pain. Initially, a benign cystic parotid lesion was suggested through imaging and FNAC. Subsequent superficial parotidectomy, guided by frozen section analysis, unexpectedly revealed non-invasive carcinoma ex pleomorphic adenoma. Postoperative radiotherapy was advised. During follow-up, Frey's syndrome manifested.

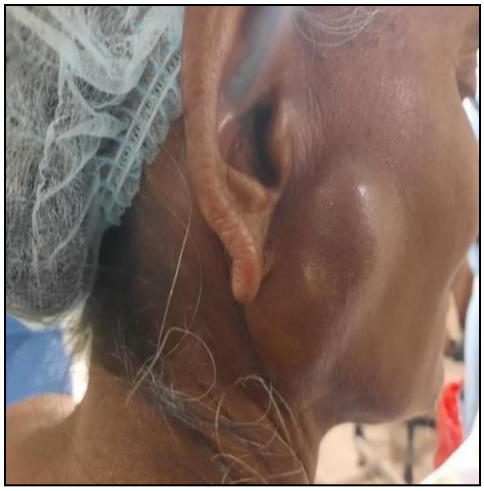

Figure 1 : Right parotid swelling.

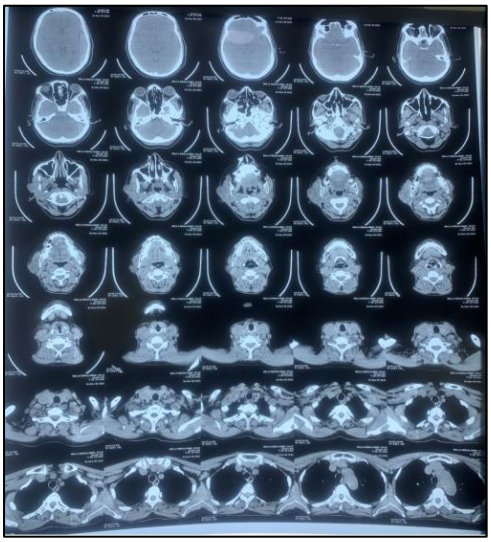

Figure 2: CT scan showing a well-defined heterogeneous density lesion, predominantly cystic withsolid components, involving the parotid gland.

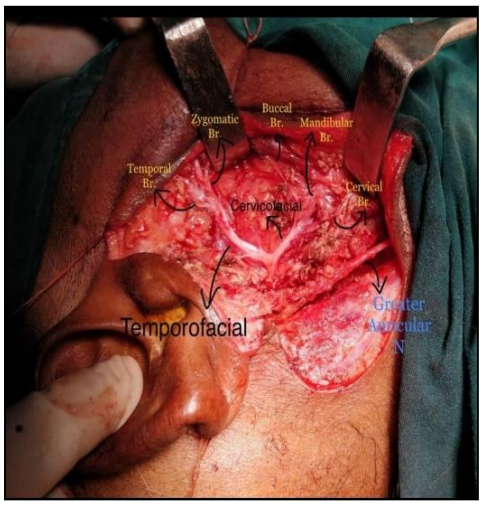

Figure 3: Right superficial parotidectomy.

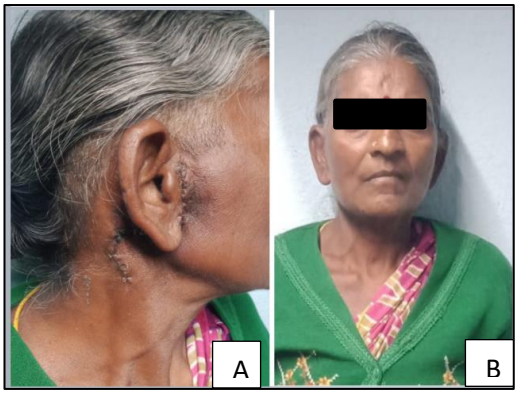

Figure 4 (A and B): Post-op follow up.

The patient, initially asymptomatic, noticed a gradually progressing swelling, reaching 3x4 cm (Figure 1). Pain emerged two weeks before presentation. No facial asymmetry or salivary-related complaints were reported. Examination revealed a well-defined, 3x4 cm swelling in the right parotid region, extending superiorly to the zygomatic arch, inferiorly to the angle of the mandible, anteriorly upto 2 cm in front of the ear lobule and posteriorly just in front of the ear lobule. USG and FNAC indicated a solid cystic lesion in the right parotid gland. CT scan confirmed a well-defined heterogeneous density lesion, predominantly cystic with solid components, involving the parotid gland (Figure 2). Superficial parotidectomy revealed benign findings on frozen section but histopathology confirmed non-invasive carcinoma ex pleomorphic adenoma (Figure 3). Postoperatively, radiotherapy was recommended and Frey's syndrome developed during follow-up (Figure 4).

DISCUSSION

Pleomorphic adenoma is the most common benign tumor affecting the salivary glands, presenting in 60–70% of cases. Sometimes, a pleomorphic adenoma may undergo malignant transformation, developing into a carcinoma ex pleomorphic adenoma or carcinosarcoma, respectively.[1,2] Pathological assessment is the gold standard for diagnosis. Prior to surgical excision, diagnosis can include fine needle aspiration cytology (FNAC), ultrasonography, computed tomography (CT) scans and magnetic resonance imaging (MRI) scans. FNAC is commonly used pre-operatively, but its sensitivity is usually low.[4]

Treatment of CXPA usually involves an ablative surgical procedure. A superficial parotidectomy is used for minimally invasive CXPA, as in our case, where the specimen was sent for a frozen section, which was suggestive of a benign salivary gland tumor and on histopathology, it was conclusive of CXPA. Total or radical parotidectomy is indicated for frankly invasive CXPA. A concomitant neck dissection might be needed if cervical lymph nodes show evidence of metastases.[9] The prognosis of CXPA depends on pathological staging parameters like the level of invasion, lymph node

involvement and local or distant metastasis. Patients with non-invasive or minimally invasive CXPA show a better prognosis.[10]

CONCLUSION

Ca ex pleomorphic adenoma is a rare yet clinically significant malignancy requiring heightened awareness. This case's atypical presentation, with a short history and contradictory diagnostic findings, highlights the diagnostic challenges associated with this entity. Accurate diagnosis relies on a combination of imaging, FNAC and histopathological assessments. Treatment involves surgical excision and adjuvant radiotherapy, with prognosis linked to pathological staging parameters. This study contributes to the understanding of Ca ex pleomorphic adenoma, emphasizing the need for vigilance in diagnosis and comprehensive therapeutic strategies.

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