CASE REPORT

Extramedullary plasmacytoma: rare neoplasm of parotid gland

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SUMMARY

A 45-year-old man presented with left-sided facial swelling. Initial excisional biopsy was done somewhere else without prior imaging and it came out to be pleomorphic adenoma; local imaging after biopsy showed mass over the left parotid gland with ipsilateral lymphadenopathy. On examination, there was a well-circumscribed mass with signs of left facial nerve palsy. He underwent left radical parotidectomy with ipsilateral modified radical neck dissection, and per operatively, it came out to be neoplastic lesion. Final histopathology was plasma cell neoplasm of the parotid gland. After surgery, the patient received radiation therapy to the left parotid region on account of uncertain surgical margins. It has been 3 years down the road that there is no evidence of local recurrence or transformation into multiple myeloma. Available evidence reveals that mere surgery for extramedullary plasmacytoma is not enough for local control whenever there is doubt of residual disease consider radiation therapy.

BACKGROUND

Solitary plasmacytoma (SP), localised lesion of the bone or soft tissue, constitutes 2%–10% of all plasma cell tumours. SP of the head and neck arising either from the bone or soft tissue is a rare entity.¹ Extramedullary plasmacytoma (EMP) most

Figure 2 Presurgery CT scan showing tumour extent in coronal views.

frequently arises in the upper aerodigestive tract. Booth *et al* in their review of 250 reported cases stated that primary EMP usually affect the head and neck region.² SP of the parotid gland is a rare condition with the first case reported in 1965.³ Definitive treatment for SP is either radiation therapy or complete surgical excision.

CASE PRESENTATION

A 45-year-old man presented with left-sided facial swelling which had been gradually increasing in size for 8 years. Considering it to be parotid swelling, he underwent superficial parotidectomy without local imaging at the same hospital and histopathology reported as pleomorphic adenoma. However, the swelling kept on increasing after surgery. Then, he consulted our hospital after a period of 1 year. On examination, there was a well-circumscribed mass in left preauricular region with signs of left facial nerve palsy. Systemic examination was unremarkable.

INVESTIGATIONS

CT scan of the head and neck was advised to see the extent of local disease and it showed a left parotid gland mass with ipsilateral lymphadenopathy (figures 1 and 2). Skeletal survey, bone marrow biopsy and serum/urine protein electrophoresis

Figure 1 Presurgery CT scan showing tumour extent in axial views.

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Figure 3 Parotid plasmacytoma—H&E stain slide with magnification $\times 40$.

with immunofixation were advised to rule out multiple myeloma (MM) and it turned out to be negative.

TREATMENT

This man underwent left parotidectomy and left-sided modified radical neck dissection based on the previous histopathological finding of pleomorphic adenoma. Intraoperatively, the left parotid lesion measured 5×5 cm involving the whole of the gland and extending superficially up to the subcutaneous tissue; left facial nerve could not be identified and multiple ipsilateral enlarged neck lymph nodes were seen. Frozen section confirmed it as a neoplastic lesion. On final histopathology, it came out to be plasma cell neoplasm on H&E stains (figures 3 and 4) with positivity for immunohistochemical markers Leukocyte common antigen (LCA), Cluster of differentiation 56 (CD56), Cluster of differentiation 138 (CD138), Multiple myeloma oncogene 1 (MUM-1) and Epithelial membrane antigen (EMA) (figure 5). However, none of the lymph node was involved. Skeletal survey, bone marrow biopsy and serum/urine protein electrophoresis with immunofixation were advised to rule out MM and all those turned out to be negative. Patient was then discussed in multidisciplinary tumour board meeting and the consensus was to offer

Figure 4 Parotid plasmacytoma—H&E stain slide with magnification $\times 400$.

Figure 5 Histopathology slide—tumour cells showing positivity for CD138 immunostain.

radiation therapy on account of piecemeal surgical resection and uncertainty of tumour's margin. Patient then consulted in a radiation oncology clinic. He was planned for a total dose of 50 Gy in 25 fractions, at a daily dose fraction of 2 Gy over 5 weeks using three-dimensional conformal radiotherapy (3DCRT) technique (figures 6–10). The patient underwent treatment according to plan and tolerated it well. He was reviewed weekly during the treatment by our team. There were no significant side effects observed except moderate skin reactions on the irradiated skin and oral mucositis at the end of treatment. He was advised to follow up after 4 weeks.

OUTCOME AND FOLLOW-UP

The patient was reviewed after 4 weeks. Dermatitis and oral mucositis were almost resolved; only mild hyperpigmentation of irradiated skin was seen. The patient had been generally doing well. He was advised to have a CT scan performed after a further 4 weeks to review the treatment response, which was negative for any residual disease. He has been on regular follow-up for 3 years with urine, serum protein electrophoresis with immunofixation, complete blood count, serum creatinine and serum

Figure 6 CT scan axial view: contouring of planning target volume, contralateral parotid gland, brain stem, spinal cord, right eye and right optic nerve on planning CT scan with different isodose levels.

Figure 7 CT scan coronal view: contouring of planning target volume and organ at risk.

calcium. Until now, there is no evidence of local recurrence or transformation into MM.

DISCUSSION

SP is characterised by a lesion having neoplastic monoclonal plasma cells in either soft tissue or bone without having systemic disease attributing to MM. So mere histopathological analysis cannot make a diagnosis of SP and MM must be excluded by relevant systemic investigations. To differentiate soft tissue plasmacytoma from the bony plasmacytoma or systemic disease, we use the following diagnostic criteria³:

- ▶ tissue biopsy shows monoclonal plasma cell histology;
- ▶ bone marrow plasma cell infiltration not exceeding 5% of all nucleated cells;
- ▶ absence of osteolytic bone lesions;
- ▶ no evidence of MM;
- ▶ absence of hypercalcaemia or renal failure;
- ▶ low serum M protein concentration.

Head and neck solitary EMPs are rare plasma cell neoplasms. In two-thirds of cases in the head and neck, EMP is located in the upper respiratory tract or oral cavity primarily in the nose, sinuses and nasopharynx region.² Solitary EMP of the parotid gland is a rare condition among all head and

Figure 8 CT scan sagittal view: contouring of planning target volume and organ at risk.

Figure 9 Anterior and posterior oblique field arrangement.

neck EMPs.⁴ The ideal treatment for EMP of the head and neck is still not clear. We know radiation plays a major role in the management of EMP; however, surgery can be used as an alternative in few selected cases.^{4–6} Radical excision is often not possible in most of the cases because of the large tumour size, involvement of surrounding structures and poor cosmetic results with surgery.⁷ An interesting question therefore arises: after surgery, do we really need adjuvant radiation for local control? We do not have a clear answer for this question yet. According to the available literature, overall local recurrence rate of salivary gland tumours is 14% (2/14 cases) with surgery alone.⁸ However, there is a risk for primary as well as nodal relapse.⁹ So, it is considerable to offer adjuvant radiation therapy to achieve local control whenever there is doubt of local residual disease. The literature reveals that patients with head and neck plasmacytoma have better prognosis than those with plasmacytoma elsewhere in the body.¹⁰ Since parotid EMP itself is a rare entity, presently, there is no literature available to guide us regarding overall survival

Figure 10 Oblique field arrangement in beam's eye view.

(OS) and recurrence-free survival. But in general, survival of EMP is always better than bone plasmacytoma.¹¹ In a review of 721 EMP cases by Alexiou *et al*, around 65% of patients had no recurrence and did not progress to MM, whereas 22% experienced recurrence, and 15% of cases evolved into MM.⁷ In another case series of 18 patients from Mayo Clinic showed that all EMP of the head and neck treated, median OS was 12.5 years with 5-year and 10-year survival rates of 88% and 55%, respectively, six patients developed MM or plasmacytoma at a distant site.¹² In addition, an analysis of the Surveillance, Epidemiology, and End Results database that included 1185 patients with EMP reported superior median survival among patients with involvement of the head and neck when compared with patients with other sites of EMP (13 vs 4 years).¹⁰ So, there can be three patterns of failure namely local recurrence, development of MM and development of new lesions without MM.¹³ Therefore long-term follow-up is necessary to detect local recurrence, or transformation into MM. In different studies available, there are variable data for conversion of EMP to MM which ranges from 8% to 36%.^{10 14}

Learning points

- ▶ All tumour cases must be discussed in the multidisciplinary tumour board meeting before embarking on treatment.
- ▶ Thorough work-up is required to make diagnosis of solitary plasmacytoma and exclude systemic disease multiple myeloma (MM).
- ▶ The established therapeutic approach is either radiation therapy or complete surgical removal of tumour. Radiation therapy alone being able to offer cure with better functional outcomes.
- ▶ Long-term follow-up is required for early detection of local recurrence or transformation into MM.

Contributors NA is primary radiation oncologist of this patient and he took this case in MDT for consensus decision. ANA and BMQ assisted in major planning of radiotherapy in PEER REVIEW meeting. SA had the idea to report this case. SA and BMQ have played a major role in writing this case report. All the authors made a significant contribution to this study.

Competing interests None declared.

Patient consent Obtained.

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