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Primary neuroendocrine tumor of the scapula

Ratna Parikh, Anuj Parikh, Sarabjeet Kaur Arneja, Praful B Desai

CASE REPORT

A 62-year-old man presented with pain and gradually increasing swelling of the right scapular region for four months. On examination, there was a bony hard swelling on the scapula with no restriction of movement. Magnetic resonance imaging (MRI) showed a destructive lesion in the spine of the scapula (Figure 1). Whole body 18-Fluoro-deoxy-glucose positron emission tomography (FDG-PET) showed a non-FDG avid lesion in the spine of the scapula, with no other metabolically active lesion identified elsewhere (Figure 2). The spine of the scapula was surgically resected with adequate margins. Histopathology revealed a neuroendocrine tumor (NET). Lesional cells showed diffused string immunopositivity for pan cytokeratin (pan CK) an epithelial cell marker and markers of neuroendocrine differentiation, chromogranin and synaptophysin (Figure 3). Upper and lower endoscopies were negative for a primary gastrointestinal NET. Postoperatively, the patient did not receive any adjuvant therapy. The patient was followed up over four years and has remained disease free, clinically and radiologically.

Figure 1: MRI 3D reconstruction showing a destructive lesion in the spine of the scapula.

Figure 2: Whole body PET-CT scan showing a non-FDG avid localized lesion in the spine of the scapula with no other metabolically active lesion identified elsewhere.

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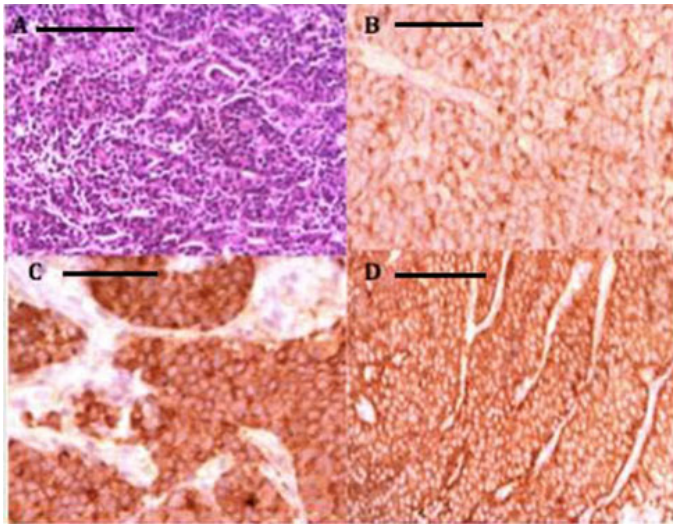

Figure 3: H&E 20×. (A) Cellular epithelial tumor, the cells are fairly monomorphic and arranged in a nested pattern. (B) Immunohistochemically the cells showed positivity for cytokeratin and (C and D) diffuse strong immunopositivity for neuroendocrine markers, chromogranin and synaptophysin. Scale bar: 0.5 mm.

DISCUSSION

Neuroendocrine tumors are a heterogeneous group of malignant tumors, arising from the enterochromaffin cells of the gut and bronchi [1]. Current data shows that NETs form 10% of all neoplasms [2]. They may vary in location, clinical presentation, tumor biology, and prognosis. The tumors can occur at any site in the neuroendocrine system. Seventy five percent of NETs arise from gastrointestinal tract, i.e., the small intestine, appendix, stomach, and rarely the colo-rectum. The lung and bronchus are the second most common sites forming 20% of all NETs [2]. The pancreatico-biliary tree, thymus, and ovary are the other known sites. Some tumors present with clinical syndromes related to peptides and amine production, or the carcinoid syndromes related to serotonin and tachykinin production from small intestinal NETs (carcinoids).

Prognosis in patients with NETs depends on its site and stage. Localized NETs in the stomach, small bowel, colon, appendix, liver, pancreas, and thymus have a median survival of 154, 111, 261, 360, 50, 136, 111 months whereas metastatic disease has a survival of 15, 56, 5, 27, 12, 24, and 40 months, respectively [3]. Patients with NETs of the appendix have the best outcomes whereas liver NETs have the worst prognosis [3].

Bone metastases arising from NETs are a well-recognized complication and carry a poor prognosis [4]. Primary skeletal NETs are extremely rare, however, it has been described in the sacrum, coccyx, cervical spine, and base of the skull [5–10]. This patient had a localized primary NET of the scapula. Extensive metastatic workup did not show any other site of NET. Though adjuvant therapy may be beneficial for skeletal metastasis from NETs, there are no studies to suggest its benefit in a localized primary skeletal NET as in this case. Such a primary lesion in the scapula remains unreported in literature.

CONCLUSION

Primary skeletal NETs are rare. Hence, it is essential to do a full body imaging using a combination of anatomic and functional imaging to rule out any additional lesions. Bone metastases and other tumors should be part of the differential diagnosis when assessing patients with NET. Primary NET arising from the scapula remains unreported in literature.

Keywords: Immunohistochemistry, Neuroendocrine tumor, Scapula

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Ratna Parikh – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Anuj Parikh – Conception of the work, Design of the work, Acquisition of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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