CASE REPORT

Unusual manifestation of a multiple myeloma in the hyoid bone

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The most common manifestation of plasma cell neoplasms is multiple myeloma. Solitary and localized tumours in the form of solitary plasmacytoma of the bone or extramedullary plasmacytoma are rare. In the late stages of multiple myeloma, bulky bone tumour infiltrates may be found which may be the primary clinical manifestation of the previously unknown malignancy. We report a case of a hyoid bone tumour with extramedullary plasma cell infiltrates in the oropharynx in multiple myeloma.

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Introduction

The typical manifestation of plasma cell tumours is multiple myeloma (MM) with multifocal infiltrations of the bone marrow and multiple osteolytic lesions of the vertebrae, skull and long bones. In later stages MM can also be seen in other bones or extramedullary tissue. This tumour is rarely found as a solitary plasmacytoma of bone (SPB) or as an extramedullary plasmacytoma (EMP).¹ We present a case of a plasma cell tumour which manifested primarily as a solitary affection of the hyoid bone and as an extramedullary infiltrate of the oropharynx, representing a late stage dissemination of a MM. The hyoid manifestation of a plasma cell tumour as described here is extremely rare, and to our knowledge only three cases of SPB of the hyoid have been published in the literature.^{2–4}

Case report

An 81-year-old male was referred to the otorhinolaryngology department because of a progressive swelling of the neck over a period of 3–4 months, a swallowing disorder and weight loss. His medical history was

irrelevant with benign prostate hyperplasia, cholecystectomy and a gunshot wound in the head which was treated neurosurgically many years ago. The patient did not have any skeletal pain, fever or recurrent infection but did suffer from night sweating.

A huge midline crossing pharyngeal tumour with lymph node metastases was suspected clinically and in ultrasound. On the contrast-enhanced CT (CECT) [LightSpeed Pro 16 (GE Medical Systems, Milwaukee, WI); 140 kV, 330 mA, slice thickness 1.25, pixel spacing 0.424×0.424 mm, pitch factor 1.375:1, 50 ml Imeron 300 (Bracco Imaging, Konstanz, Germany), antecubital vein, flow 3 ml s^{-1} , delay 50 s] there was some asymmetry detected with an accentuated right pharyngeal tonsil (Figure 1a,b), a huge tumour of the hyoid bone (Figure 1c–f) and an asymmetry of the piriform recess (Figure 1f). The tumour and hyoid bone destruction could also be seen on a lateral radiograph of the neck (Figure 2a,b). The bone was nearly completely destroyed; the left cornu was partially spared, funnel shaped and expanded with calcification at the margin of the tumour.

The soft tissue mass exhibited vigorous, slightly inhomogeneous contrast enhancement up to 100 HU and measured 67×32 mm in the axial plane. The mass was sharply delineated without obvious infiltration into the neighbouring anatomical structures; these were considerably dislocated, especially the pharynx. The CT scan revealed some small lymph nodes without malignant

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Figure 1 Contrast enhanced axial CT scans in the craniocaudal direction. (a,b) Scans at the level of the tonsils reveal the asymmetry with a bulky right tonsil (white arrow). (c–e) Scans at the level of the hyoid bone reveal a sharply delineated mass destroying the hyoid bone (white arrows in c and d). (f) Scan at the level of the piriform recess. The piriform recess on the right is compressed; on the left it is normally configured (white arrow)

suspicion. Under these conditions the possibility of a primary non-metastatic affection of the hyoid was hypothesized.

A panendoscopy carried out in the otorhinolaryngology department revealed intact mucosa in the respiratory and upper digestive tracts, compression of the right piriform recess and some indurations of the right pharyngeal tonsil. Multiple biopsies from the suspicious tonsil, the right piriform region and the bilateral vallecular region were obtained. The histological evaluation showed EMP in the left vallecula with immunohistochemical positivity for VS 38 C (marker of plasma cells) and CD 138 (CD 138 is expressed in plasma cells of bone marrow and differentiates them from B-cell precursors which are CD 138 negative), negativity for CD 20 (CD 20 is a marker of B-cells in peripheral blood and lymphoid tissue), an intracellular over-expression of κ -chains and nearly negative λ -chains.

The haemato-oncological examination revealed osteoporosis with radiological detection of suspicious focal skeletal osteolytic lesions, hypercalcaemia (2.94 mmol l^{-1} , range of normal values $2.25\text{--}2.65 \text{ mmol l}^{-1}$) and a haemoglobin level of 9.6 mmol l^{-1} (normal range $8.6\text{--}12.0 \text{ mmol l}^{-1}$). Renal function was normal with a creatinin value of $79 \mu\text{mol l}^{-1}$ (normal range $<102 \mu\text{mol l}^{-1}$).

Pathological proteins could not be detected in the urine or serum. The bone marrow biopsy revealed 30% infiltration of plasma cell myeloma. The disease was classified as non-secretory MM stage IIIA according to Durie and Salmon.⁵ Because of the systemic disease, chemotherapy with bendamustin and prednisolon together with antihypercalcaemic therapy with pamidronate was initiated.

Discussion

Plasma cell neoplasms constitute approximately 10–15% of haematopoietic tumours and about 1% of all malignancies with the frequency of 4–6 cases per 100 000 a year. It is a malignancy which occurs in older age with a slight predominance in males.¹

Plasma cell malignancies belong to the low grade B-cell non-Hodgkin lymphomas. In this tumour the monoclonal proliferating malignant plasma cells accumulate in the red bone marrow⁶ or in the submucosa.⁷ The most common manifestation (approximately 90% of cases) is multifocal infiltration of the haematopoietic bone marrow. The remaining are localised forms: approximately 5% of cases are SPB and are reported in the

Figure 2 (a) Full view lateral cervical spine radiograph. (b) Detail of the lateral radiograph of the cervical spine demonstrates a huge soft-tissue mass and destruction of the hyoid bone

vertebra, ribs, skull base and, rarely, other bones^{8–14} and approximately 5% are EMP, with the most frequent site being the upper respiratory tract.^{1,8,15,16} The localized forms exhibit strong predominance in males.

The diagnosis of a localized form of plasma cell tumour assumes the exclusion of MM,¹⁷ which means (a) the bone marrow biopsy taken from a site remote from the tumour should be normal with no malignant plasma cells or less than 10% of plasma cells in the specimen and (b) no anaemia should be present.¹⁴ However, the progression of a solitary plasmacytoma into MM is known and, even after surgery and/or radiotherapy, almost 15–39% are in EMP and 50–84% are in SPB.^{14–19} Corwin and Lindberg¹⁴ reported progression from solitary forms to MM within 24 months. They suppose a 3 year disease-free interval as a criterion for the distinction of SPB from EMP. The relationship between MM and SPB is not yet clear; two contradictory opinions can be found in the literature—SPB is solely a manifestation of a MM²⁰ or SPB is a true precursor of subsequently developing MM.²¹

The tendency of SPB to progress to MM may be owing to an occult MM at the time when SPB manifests.⁸

Clinical manifestations of MM are: accelerated turnover of the bone matrix, such as osteoporosis, osteolytic lesions, skeletal pain, pathological fractures and hypercalcaemia caused by osteoclast activation; destruction of the haematopoietic bone marrow such as anaemia, leucopenia and thrombocytopenia; and specific findings of

paraprotein secreted by tumour plasma cells which form amyloid deposits in the kidneys with resulting renal failure or amyloid depositions in other organs. The immunodeficiency manifests as recurrent infections. In late stages the extramedullary plasma cell infiltration of other organs is possible but rare,²² either in the form of bulky lesions or diffuse infiltration. The evolution of bulky plasma cell infiltration in bones is also possible but mainly occurs in the late stages of MM. The primary manifestation of previously unknown MM as a local skeletal lesion is unusual.^{23,24}

In cases of SPB or EMP, local symptoms such as swelling, pain, obstruction of the airways or bleeding are dominant.²⁵

In our patient the leading symptoms of neck swelling and swallowing disorders were caused by the expansion of the hyoid mass and dislocation of the pharynx, larynx and upper oesophagus.

The radiological manifestation of the hyoid mass corresponds with other published cases.^{2,3} The CT revealed a sharply circumscribed expansive mass without infiltration of the collateral structures. The hyoid was destroyed and the rest of the cortical bone was expanded into a funnel shape, as reported in cases of solitary plasmacytoma of the hyoid and the rib. The sclerosis was absent. The mass exhibited vigorous enhancement after the intravenous application of contrast material, as described in CT of EMPs²⁵ and as is usually found in the contrast-enhanced

MRI of MM lesions.²⁶ The reports on enhancement of MM and plasmacytoma in CT are rare because of the known risk of renal failure when iodinated contrast material is administered but an analogous enhancement pattern on MRI can be expected. Only cases in which plasma cell proliferation is not known or not suspected are examined in CT with contrast enhancement, as it was in our case.

In the biopsy specimen of the right tonsil that appeared accentuated in the CT scan and indurated in panendoscopy, only stromal swelling without malignancy could be identified pathologically; the localization of plasma cell infiltrates deeper in the tissue could be hypothesized. However, the left vallecula with plasma cell infiltrates showed no detectable tumour or enhancement in CT owing to a limited number of tumour cells. Marked osteoporosis in the vertebrae was noted.

In the patient the medullary and extramedullary plasma cell infiltrates could be finally diagnosed. The concomitant occurrence of infiltration of both compartments rather excludes the diagnosis of solitary EMP according to the criterion mentioned above.

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