European Annals of Otorhinolaryngology, Head and Neck diseases 138 (2021) 301–302

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What is your diagnosis?

Progressive facial deformity

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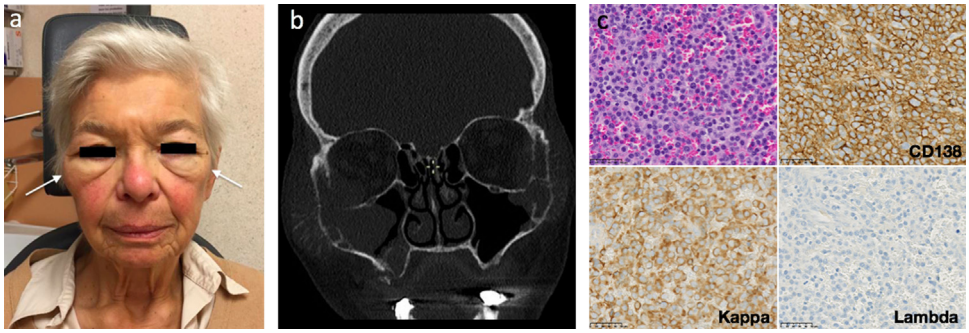

Fig. 1. a: facial masses in an 81-year-old woman; b: CT scan of the facial bones, coronal section; c: histological and immunohistochemical examinations.

1. Description

An 81-year-old woman consulted for painless, non-inflammatory masses on the forehead, zygomatic arches and right maxilla (Fig. 1a) gradually enlarging for more than a year. She reported asthenia, weight loss of 2 kg and frontal headache. She did not present any notable history. The rest of the otorhinolaryngological examination was normal.

CT scan of the head and neck showed multiple poorly demarcated osteolytic lesions in the mandible, left arytenoid, skull base, manubrium sterni and clavicles and osteolytic lesions with "sun-ray" radial periosteal spicules in the zygomaticomaxillary complex and frontal bone (Fig. 1b). PET scan and MRI confirmed these lesions, and also demonstrated lesions of the ribs, spine and limbs, with no soft tissue lesions.

Complete blood count was normal. Serum protein electrophoresis revealed a monoclonal IgG peak of 30.8 g/L with a large quantity of Kappa light chains on immunofixation (401.1 mg/L) and a Kappa/Lambda light chain ratio of 222.83 (> 1.65). Urinalysis revealed Bence-Jones proteinuria.

This work-up was completed by bone marrow aspiration, bone marrow biopsy, and bone biopsies of the right maxilla (Fig. 1c).

What is your diagnosis?

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https://doi.org/10.1016/j.anorl.2020.10.011

2. Answer

Several diagnoses must be considered, such as bone metastases from cancer of the kidney, thyroid, colon, rectum and melanoma that can give rise to metastases in the facial bones. Other tumours, such as osteosarcoma, Ewing's sarcoma and chondrosarcoma may present with this "sunray" radiological appearance, but appeared to be unlikely in this case [1]. Bone marrow examination showed a monoclonal IgG Kappa plasma cell population accounting for 10% of cells. Biopsies of the maxillary lesion confirmed a proliferation of plasma cells expressing CD138 and mostly Kappa chains (Fig. 1c). Based on the criteria defined by the International Myeloma Working Group in 2014 [2], the diagnosis adopted was that of multiple solitary bone plasmacytomas in a context of IgG Kappa multiple myeloma (MM).

Plasmacytoma is a monoclonal plasma cell infiltration arising in bone or soft tissue. Two entities have been described, solitary plasmacytoma and multiple solitary plasmacytomas. They are characterized by the absence of bone marrow involvement or only minimal bone marrow involvement (plasma cell infiltration < 10%) [3,4] At the time of diagnosis, this patient had already progressed to multiple myeloma. The 10-year rate of progression to multiple myeloma after the initial diagnosis of bone plasmacytoma is estimated to be 50% [3,4].

As the diagnosis of plasmacytoma is histological, there are no specific radiological criteria. In MM, radiographic bone lesions mainly consist of osteolytic lesions and more rarely osteoporotic or osteosclerotic lesions [1]. The unusual feature of this patient was the presence of four bone plasmacytomas in the facial bones with fairly similar radiological features, comprising osteolysis and osteosclerosis in the form of bone spicules ("sunray" image) (Fig. 1a and b). Primary osteosclerosis is rare in MM and may present as diffuse or focal osteosclerosis, bone spicules or an osteosclerotic reaction surrounding an osteolytic lesion [1]. The presence of a histologically confirmed bone plasmacytoma with a radiological signs of osteosclerosis with bone spicules appears to be a rare association, as only a few cases have been reported in the literature [5].

This patient was treated by chemotherapy for her MM, as the presence of plasmacytomas did not alter the treatment of MM. The treatment of solitary bone plasmacytoma consists of radiotherapy and/or surgical resection [3].

Disclosure of interest

The authors declare that they have no competing interest.

Acknowledgements

The authors would like to thank Professor Alessandra CAMBONI of the Saint-Luc University Clinics Pathology Department in Brussels for the histological diagnosis and for providing the histological and immunohistochemical illustrations.

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