Solitary extra-medullary plasmacytoma of the true vocal cord

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To the Editor: Solitary plasmacytoma is an uncommon plasma cell neoplasm that accounts for 5% to 10% of all plasma cell neoplasms. It is divided into two categories based on location: solitary bone plasmacytoma (SBP) which occurs in bone and solitary extra-medullary plasmacytoma (EMP) in soft tissue. SBP is 40% higher than EMP. Although EMP can arise throughout the body, 90% arise in the head and neck, especially the nasal cavity, paranasal sinuses, and pharynx.[1] Nevertheless, EMP of the larynx is a rare situation, accounting for 0.04% to 0.19% of all malignant laryngeal lesions. And the variant appearances of laryngeal EMP make the accurate diagnosis quite difficult without the final pathologic result. Herein, we reported a case of EMP that was misdiagnosed as a vocal cord polyp.

After catching a cold, a 42-year-old male experienced acute onset of progressive hoarseness and persistent cough. His primary physician prescribed a course of antibiotics for laryngitis which turned out ineffective. His symptoms persisted for about 6 months and a vocal fold lesion was suspected. So he was admitted to the Peking University Third Hospital. The patient denied pharyngalgia, dysphagia, dyspnea, fever, or weight loss in the past 6 months. He had smoked for more than 20 years, two packs a day. Indirect and flexible laryngoscope revealed a pink exophytic mass with a narrow base on the middle third of right vocal fold. The neoplasm had a smooth surface with a diameter of about 10 mm [Figure 1]. The movement of the vocal fold was normal, and no abnormality was found in the nasopharynx or oropharynx, except for some non-specific chronic inflammation signs. No superficial lymph node was palpable. Taken together all the clinical information, a benign lesion such as polyp was presumed. So without a biopsy, a direct suspension laryngoscopy was performed under general anesthesia. The lesion was resected with CO_2 laser following the phonomicrosurgery principle.

The pathological examination of the lesion demonstrated diffused and dense infiltration of plasma cells, and immunohistochemical tests indicated plasmacytoma as the correct diagnosis. A complete work-up for multiple myeloma (MM) was then carried out, revealing normal serum electrophoresis, serum immunoglobulins, and β2-microglobulin levels. Complete blood count (CBC), serum calcium, uric acid, and renal function tests were normal. Bone marrow aspirate and biopsy of the right iliac crest showed normal hematopoiesis with less than 1% plasma cells. Skeletal survey, including X-rays of the long bones and spine, was normal. A whole-body positron emission tomography scan showed no evidence of metastatic disease. Urine examination for light chains indicated slightly increased kappa light chain and lambda chain. The diagnosis of laryngeal EMP was made after all those efforts.

The latest guidelines for solitary plasmacytoma recommend radiation therapy to the involved field as primary therapy, and surgery is done as an adjunction before radiation if necessary.[2] But the patient declined radiation therapy, and was under close follow-up. Every 6 months, a flexible laryngoscopy was performed to monitor local recurrence, while CBC and urinalysis were carried out for systemic recurrence monitoring. At 5-year follow-up, the patient remained asymptomatic. Laryngoscopy and laboratory tests showed no signs of local or systemic recurrence.

EMP represents less than 1% of tumors in the head and neck, most commonly in the upper respiratory tract and generally in male patients in their 40s to 70s. Laryngeal EMP, which comprises 4.5% to 18.0% of all EMP, is rare but has also been reported more and more recently.[3] The major clinical manifestations are hoarseness, others include dyspnoea, dysphagia, stridor, sensation of a foreign body, hemoptysis, and dry cough, which are dependent on the site and size of the lesion.[3] Some patients complain of a neck mass on their first visit to an otolaryngologist, and a few can be asymptomatic. Different appearance of laryngeal EMP has been described. Lesions can be as small as 0.3 cm or as large as 3.0 cm, forming a solitary granulation polyp or diffused thickening of mucosa or a mass in the larynx. Supraglottic region is

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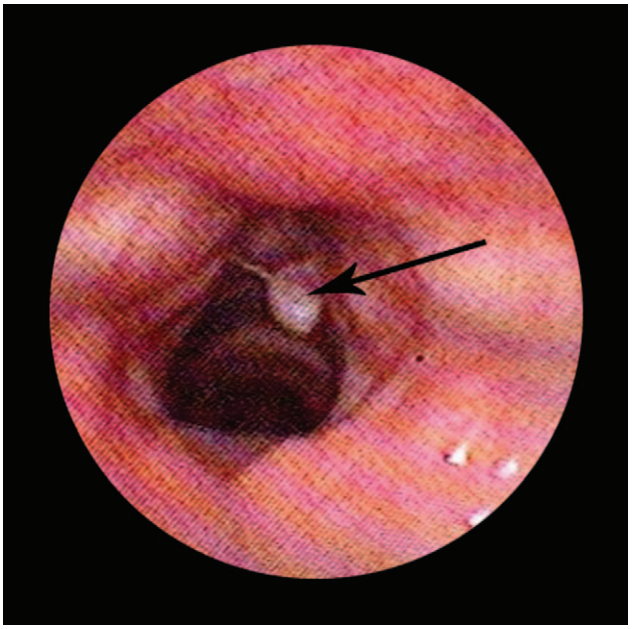

Figure 1: Endoscopic result of a 42-year-old male with solitary extra-medullary plasmacytoma before surgery.

the most commonly affected site, followed by glottic region and subglottic region.[1,3]

As for our case, the lesion was just at the glottis and was so small that the patient complained nothing but hoarseness. Considering the smooth and polypoid appearance, it was misdiagnosed and treated the same way as what we usually do to a vocal cord polyp. From this case, we think it is important to emphasize that plasmacytoma is a special malignant entity that could imitate benign lesions. The correct diagnosis can be challenging from laryngoscopy.

There have been numerous advances in the treatment of MM during the past few years, but the treatment paradigm for solitary plasmacytoma has not changed significantly. Surgery combined with radiotherapy is considered superior than uni-modality in preventing local recurrence, but radiotherapy is still the mainstay of plasmacytoma.[1] Guidelines from the International Lymphoma Radiation Oncology Group recommend that a definitive surgical excision alone can be considered as acceptable treatment only for small tumors in anatomic locations where clear margins are attained with minimal morbidity.[2] Cutaneous lesion and solitary lung lesion are taken as examples where surgical excision alone is acceptable. In this case, when the final diagnosis first came out, we strongly advised the patient to receive radiotherapy, but the patient refused for fear of the adverse effects. Considering the clear margin was not so sure, we followed this patient closely. Fortunately, examination of the patient still showed no sign of local recurrence or progression to MM at 5-year's follow-up. We thought this might imply a clear margin by CO_2 laser for the sealing effect could be 2 mm deeper.[4]

On the other hand, according to the literatures, factors related with risk of EMP local relapse and myeloma progression include not only the type of treatment, but also the site of disease, age of the patient, size of the lesion, and bony erosion. EMP of non-sinonasal region has a better prognosis than sinonasal lesions. Patients over 65 years, lesions larger than 5 cm or with bony erosion had a worse prognosis.[1] Our patient was a 45-year old male with a restricted lesion in the larynx, with no evidence of bony erosion, which meant that he did not belong to the high-risk group. Despite the relatively good prognosis of EMP, there still have been 10% to 15% of EMP cases reported to progress to MM.[1] So life-long follow-up and surveillance are essential for this patient, including serial and frequent blood and urine tests.

In summary, laryngeal EMP is a rare disease with various confusing appearance. Radiation therapy should be prescribed whenever possible and close follow-up is needed for patients with high risks of recurrence and myeloma progression.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

None.

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