Long-Term Follow-Up of Two Cases of Primary Pulmonary Plasmacytoma

Ioannis Bougioukas*, Mohamad Shadi Alzin, Elke Eltze, Ralf Seipelt and Hanno Huwer

1Department of Cardiothoracic Surgery, Voelklingen Heart Centre, Germany
2Institute for Pathology, Saarbruecken-Rastpfuhl, Germany

Abstract

Primary pulmonary plasmacytoma is a rare type of extramedullary plasmacytoma and can potentially progress to multiple myeloma. Certain variability in the therapeutic management of this condition exists because of its rarity. Its presentation as a single pulmonary mass renders both surgery and radiotherapy as attractive therapeutic options. We present the 10-year follow-up report of two cases of primary pulmonary plasmacytoma, both managed with surgical sublobar resection solely. In the first case, the follow-up was completely uneventful. In the second case, a new pulmonary mass was demonstrated seven years after the resection; however biopsy of this mass excluded both malignancy and disease progression. The actual follow-up of this patient showed no further change of the radiological finding. The possibility of existence of an amyloidoma on the old surgical scar tissue has been considered. Looking at the scarce literature, we conclude that complete surgical resection accompanied by regular aftercare is a rational therapeutic concept for primary pulmonary plasmacytoma. Performance of sublobar resection instead of a more extensive lobectomy is sufficient minimizing operative risk and hospital stay.

Keywords: Plasmacytoma; Amyloid; Lung

Introduction

A plasmacytoma is a mass of neoplastic monoclonal plasma cells found either in bone or soft tissue (extramedullary) [1]. Extramedullary plasmacytoma (EMP) comprise 3% to 5% of all plasma cell neoplasms and are mostly involving the upper respiratory tract [2-4]. Primary pulmonary plasmacytoma (PPP) is a very rare type of EMP, with only few reports found in the literature [5-7]. The diagnosis of PPP is typically discovered by incident, often after a surgical resection. The rarity of this condition allows certain variability in its therapeutic approach and although guidelines for treatment of EMP do exist, these cannot strictly apply on PPP cases [8]. Moreover, the need for adjuvant therapy for PPP remains totally controversial. On the other hand, the risk for later progression to multiple myeloma always exists, rendering the long-term follow-up of such patients totally crucial [9]. In this report, we present the long-term follow-up results of two cases of PPP managed with surgical resection solely [10].

Case Presentation

We report two female patients who underwent a pulmonary resection due to PPP 10 years ago at our institute. Case A was then a 20-year-old woman with a CT-scan finding of a 2.5 cm thin-walled cyst, containing a solid tissue formation, located in the left upper lung lobe. A diagnostic thoracotomy was performed and the pulmonary mass was completely resected by wedge resection. Case B was then a 68-year-old woman with a radiological finding of an 1.5 cm solid nodule with scalloped and spiculated borders in the right lower lobe. A wedge resection was performed. Frozen sections of the tumours ruled out lung cancer and histological analysis yielded plasmacytoma in both cases. Further immunohistochemical stains showed that the tumor cell cytoplasm in case A stained for immunoglobulin G and kappa (κ) light chains, whereas in case B it stained for immunoglobulin A and lambda light (λ) chains. Interestingly, in the immediate vicinity of the infiltrates of monomorphic plasma cells, homogenic amyloid deposits could be seen in both cases. The plasmacytoma surrounded depositions of amyloid in the middle of the parenchymal mass. The exact findings have been already described [10]. Postoperatively multiple myeloma was ruled out by radiological examination of the axial skeleton and bone marrow cytology. Monoclonal paraprotein detection in urine and serum was negative in both cases. The 10-year follow-up of patient A showed no progression or relapse. No adjuvant therapy was applied and the patient remains in excellent health.
physical condition without any respiratory impairment. Patient B was diagnosed with rheumatic arthritis a couple of years after the thoracic procedure, for which she went on methotrexate. Five-year follow-up showed no relapse and the patient was in a good physical condition, whereas two years later she was admitted to the hospital for investigation of a collapse. CT-scan showed then a 4 cm solid mass in the remaining right lower lung lobe. A bronchoscopy with transbronchial biopsy of the mass was performed and malignancy was excluded. Examination of the sample did not reveal any plasma cells. The possibility of amyloid deposition on the old lung tissue scar was speculated, taking into consideration the intra operative primary biopsy. Moreover, similar depositions on the heart were demonstrated in echocardiography. Nevertheless, no adjuvant therapy was required, current follow-up showed no progress of the radiologic pulmonary finding and the patient is still doing fine without any respiratory compromise 10 years after the pulmonary wedge resection.

Discussion

Multiple myeloma is a systemic monoclonal plasma cell neoplasia, involving primarily the bone marrow [11]. EMP is a plasma cell tumor which arises outside the bone marrow and is generally observed in the head and neck, commonly involving the nasal cavity and nasopharynx [1,2,12,13]. An EMP located in the lungs is referred to as a primary or solitary pulmonary plasmacytoma (PPP) and its incidence is very rare [7,14]. Few reports of verified PPP can be found in the literature [5-7,9,14]. In these reports, there was no specific sexual predilection with a male to female ratio (1.4:1) and a median age of 55 years in contrast to other EMPs which have a male to female ratio of 3:1 to 5:1 [1]. Multiple myeloma can also involve the lung and in order to differentiate it from EMP, bone marrow examination is required. EMP patients should have bone marrow plasma cell infiltration lower than 5% of all nucleated cells and a normal skeletal survey [5]. Serum M protein or Bence-Jones light chains in urine may be negative in EMP [15]. The 10-year overall survival rate of EMP is reported to be around 70% [12,16]. EMP can eventually progress to MM; in a large review of over 700 EMP cases, after treatment, 15% of the cases evolved into MM and about 65% had no recurrence [17]. In comparison to solitary plasmacytoma of the bone, prognosis of EMP is significantly better with lower risk of progression to MM [18]. Radiotherapy plays a significant role in the treatment of EMP, since in most cases the tumors are located in the head and neck and a complete surgical removal could be cumbersome and somewhat mutilating [19,20]. Guidelines on the management of solitary extramduillary plasmacytoma recommend treatment of a solitary EMP by radiotherapy encompassing the primary tumor with a margin of at least 2 cm, but again these guidelines mainly refer to EMPs located in the head and neck [8]. On the other hand, in cases where an EMP is located in other areas, as for example in the lungs, surgery should be considered, especially if tumor-free resection could be achieved. In such cases adjuvant radiotherapy is probably not required [17]. Adjuvant chemotherapy seems to have no particular beneficial effect on control or progression to MM [21]. In other words, when a margin-free resection could be achieved, especially without implementing amputating surgery, surgical resection could be considered as the first-line treatment. As primary pulmonary plasmacytoma is a rare medical condition, a scarcity of literature reports about the overall survival can be found. Koss et al reported 5 cases of PPP; two of them survived over 20 years and died of non-tumor related causes. Interestingly, three of the five patients underwent a pneumonectomy as the tumor was located centrally in hilum [14]. Montero et al. reported 2 cases with progression-free follow-up reaching 10 and 15 years respectively [22]. These are the most recent literature reviews with longer follow-ups, exceeding ten years. Several other sporadic reports present relatively short-term results ranging from 6 months to 1.5 years [6,7,23,24]. In most cases, no disease progression was noticed, but on the other hand this does not preclude a possible progression at a later phase. According to Edelstein and colleagues, who reported on earlier literature reviews going back to 1959, 9 of 22 patients with PPP developed multiple myeloma, often 10 to 12 years after the initial diagnosis [9]. Addressing the question, whether surgery as sole therapy is sufficient for cases of PPP, it is imperative that safe conclusions can be drawn only with long-term follow-ups. Analysing the previous 4 cases with longer follow-ups, two cases were treated with surgery alone, one only with radiotherapy and one patient was managed with surgery and adjuvant radiotherapy [14,22]. Looking at our two cases, the diagnosis of plasmacytoma was granted postoperatively. In both cases and after consultation with the local tumor board, no additional therapy was implemented. Follow-up of both cases reached ten years. Once again, thorough examination of both patients after the initial finding of plasmacytoma excluded the diagnosis of MM. In the case of the younger female patient A, no relapse and no progression to MM were noted. In the case of the elderly female patient B, today almost 80 years-old, a small mass was demonstrated in the computer tomography of the thorax, seven years after the surgical wedge...
resection of segments 8 and 9 of the right lower lobe. A biopsy was performed via bronchoscopy and malignancy was excluded. The patient denied further invasive investigation of this finding. The actual ten year follow-up of this patient showed no disease progression, a fact that per se rules out any malignant advance. In the primary specimen examination of the elderly patient, amyloid deposits were found near the plasma cell tumor. A case of co-existence of plasmacytoma with amyloid has already been described in the literature [25] and more interestingly, cutaneous plasmacytomas associated with local deposition of amyloid have been described in dogs [26]. A possible interpretation of the later evolved mass in this patient could be an amyloidoida, but again lack of a more invasive biopsy of the mass renders this theory presumptive. Both of our cases were handled with sublobar resection avoiding lobectomy. This of course can be explained by the fact that in both patients the parenchymal mass was located relatively peripherally, precluding the need for a more extended resection. Additionally, rapid frozen section in both cases was not compatible with a primary lung malignant tumor, rendering these atypical lung resections as reasonable. Avoiding a lobectomy or even a pneumonectomy reduces both morbidity and hospital stay of these atypical lung resections as reasonable. Avoiding a lobectomy or even a pneumonectomy reduces both morbidity and hospital stay of these atypical lung resections as reasonable. Avoiding a lobectomy or even a pneumonectomy reduces both morbidity and hospital stay of these atypical lung resections as reasonable. Avoiding a lobectomy or even a pneumonectomy reduces both morbidity and hospital stay of these atypical lung resections as reasonable. Avoiding a lobectomy or even a pneumonectomy reduces both morbidity and hospital stay of these atypical lung resections as reasonable. Avoiding a lobectomy or even a pneumonectomy reduces both morbidity and hospital stay of these atypical lung resections as reasonable. Avoiding a lobectomy or even a pneumonectomy reduces both morbidity and hospital stay of these atypical lung resections as reasonable.

References


