1. Introduction

Paragangliomas of the posterior mediastinum are rare neuroendocrine tumors that arise from the aortosympathetic paranglia [1]. Most of the functional mediastinal paragangliomas were noted during the investigation of hypertension or other symptoms secondary to catecholamine excess. Non-functional mediastinal paragangliomas are noted incidentally and the diagnosis is confirmed postoperatively. Mediastinal paragangliomas are hypervascular tumors and many of them directly invade or firmly adhere to the adjacent mediastinal organs, such as the heart, great vessels, the trachea and the spine [1–3]. Therefore, many authors reported their experience of massive bleeding during tumor biopsy or tumor excision. We present a case of non-functional paraganglioma of posterior mediastinum that experienced massive bleeding during the first video-assisted thoracoscopic surgery in a local hospital. The tumor was completely excised by lateral thoracotomy with bipolar electrocautery during the second operation in our hospital.

2. Case report

A previously healthy 49-year-old female was admitted to a local hospital with a history of chronic right backache for 2 years. She did not report any other symptoms, such as headache, palpitations or sweating, and had no other significant past medical or travel history. Physical examination and laboratory data were all normal. Chest computed tomography (CT) revealed a 5 × 3 × 3 cm posterior mediastinal tumor at the T6–T9 vertebral level of the right paraspinal region (Fig. 1). With a presumptive diagnosis of neurogenic tumor, she underwent video-assisted thoracoscopic surgery (VATS) for tumor excision. However, a hypervascular tumor was noted under thoracoscopy, and massive mediastinal hemorrhaging occurred just after dissection of the posterior mediastinal tumor, which required emergent mini-thoracotomy for hemostasis. Perioperative blood loss was 700 ml. Tissue biopsy was abandoned during the operation due to the ease of bleeding. The patient was then referred to our hospital for further management.

A chest magnetic resonance image (MRI) showed no spinal cord compression or direct vertebral invasion (Fig. 1). Posterior mediastinal tumor excision was performed by extending the previous right mini-thoracotomy wound, 1 week after previous surgery. Multiple prominent feeding vessels, arising from intercostal arteries, were noted. Bleeding was not easily controlled by unipolar electrocautery. We used bipolar electrocautery instead to maintain hemostasis, and the tumor was successfully removed. Perioperative blood loss was 250 ml. No remarkable blood pressure changes occurred during surgery. Microscopically, the tumor consisted of cells arranged in nests (‘zellballen’) with a vascular stroma (Fig. 2a). An elaborate vascular network surrounded the nests of cells. Some nuclei were large and hyperchromatic. Marked hemorrhagic necrosis, stemming from previous surgery, was noted. Tumor cells were strongly immunoreactive to chromogranin (Fig. 2b) and synaptophysin but negative for cytokeratin, confirming
paragangioma. Postoperative course was uneventful and the patient was discharged on postoperative day 4.

3. Comment

Pheochromocytomas are hypervascular tumors that arise from the chromaffin cells of the sympathetic nervous system [1]. Pheochromocytomas are referred as tumors arising from the adrenal medulla. When the tumors arise from the chromaffin cells at another site, they are referred to paragangliomas or chemodectomas [1]. Most extraadrenal paragangliomas locate in the abdomen [1]. Mediastinum is a less commonly involved site of extraadrenal paragangliomas. Functional mediastinal paragangliomas are often discovered during the surveillance of hypertension and other symptoms secondary to catecholamine secretion, such as headache, palpitation, sweats, and tremor [1–3]. Non-functional mediastinal paragangliomas are asymptomatic and usually found incidentally. The diagnosis of non-functional mediastinal paragangliomas are generally confirmed by the histopathologic study after operation. The behavior of mediastinal paraganglioma is aggressive. The distant metastasis rate was reported as 19.5% and 26.6% in two review studies [3, 4]. Survival is also significantly increased in the complete resection group than in partial excision and adjuvant treatment (84.6% vs. 50.0%; 125.7±18.7 vs. 71.5±13.8 months, P<0.01) [3]. Therefore, complete surgical resection is the treatment of choice. Because of their hypervascular characters and location close to the great vessels, trachea, spine and heart, complete resection of mediastinal paragangliomas remains a surgical challenge [1–3]. Physicians at the Mayo Clinic reported their 24 years experience in 14 mediastinal paraganglioma patients. The complete resection rate was 76.9% and the mortality rate was 7.1% [2].

For functional paragangliomas, alpha-adrenergic blockade should be prescribed preoperatively to avoid perioperative hypertensive crisis. Beta-adrenergic blockade and calcium channel blocker can be used with alpha-adrenergic blockade for uncontrolled hypertension [1, 2, 5, 6]. If the diagnosis is confirmed preoperatively, angiography for evaluating the tumor vascular supply and embolization to reduce perioperative bleeding is recommended [7, 8].

Some authors reported their experience, such as in our case, that massive bleeding was noted during the biopsy or operation [2, 7–10]. Since preoperative diagnosis is difficult, VATS is usually the initial approach for a well-defined mediastinal tumor. However, the open method seems better for limiting bleeding. If the bleeding becomes difficult to control, the tumor excision should be postponed. Preoperative angiography for embolization, to reduce perioperative bleeding, is recommended before the second surgery to complete tumor excision [7, 8]. Cardiopulmonary bypass may be necessary if the tumor location is close to great vessels or heart, especially for those anterior or middle mediastinal paragangliomas [2, 3, 6]. The tumor can be completely excised by lateral thoracotomy or sternotomy with careful bleeding control. In conclusion, non-functional paraganglioma should be regarded as a differential diagnosis if the surgeon encounters an unexpected hypervascular tumor of the mediastinum.

References


