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Peripheral primitive neuroectodermal tumors: a rare case report

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Abstract We aimed to explore the diagnosis and treatment of peripheral primitive neuroectodermal tumors (pPNETs). We retrospectively analyzed the diagnosis and treatment process of a patient who was diagnosed with pPNETs by pathology. This case was a man with soft masses arising from the left chest wall near the armpit and left supraclavicular of a 47-year-old man. The patient mainly presented with the masses which increasing gradually with obvious pain. Needle biopsy showed that they were both metastatic adenocarcinoma. Ultrasonography B revealed blood flow of these two low density placeholders can be seen in the signal, not oppression axillary and vein. Radical resection of the masses were performed. Histopathologic study and immunohistochemistry (IHC) confirmed the masses to be peripheral primitive neuroectodermal tumors. pPNETs is a rare malignant small round cell tumor. CT and MRI examination can estimate the resectability of the tumor; Ultrasound B can make sure its inside blood supply and the positional relationship between the mass and the surrounding vasculature. The diagnosis of pPNETs is based primarily on histopathologic study and IHC, especially those with the characteristics of the Homer-Wright and neuroendocrine markers. Radical resection of the tumor is the most effective therapeutic method. The effect of adjuvant chemo-radiation is worth affirmation. Autologous stem cell rescue besides adjuvant chemotherapy has been associated with prolonged survival.

Key words peripheral primitive neuroectodermal tumors; immunohistochemistry; adjuvant chemoradiation

Primitive neuroectodermal tumors (PNETs) is a rare malignant small round cell tumor which was first reported by Hart [1] in 1973. According to different invasion sites, PNET can be divided into centralprimitive neuroeetodermal tumors (cPNETs) and peripheral primitive neuroectodermal tumors (pPNETs) [2]. It used to appear in children and adolescents. It usually can be found in soft tissue and bone. Up to now, there has not been a rare case reported two masses in one people, especially one was in the left chest wall near the armpit and another was in left supraclavicular. In the present study, one case of primitive neuroectodermal tumors was reported in this paper, and the clinical features as well as treatment approaches of similar cases in literatures were discussed.

Case report

A 47-year-old man was admitted to our hospital because of finding two masses in the left chest wall near the armpit and left supraclavicular for more than one year, recently the masses increased gradually with obvious pain. In the process of the disease, he also had tinnitus, dizziness, diplopia, and morning epostaxis sometimes.

Physical examination revealed that one was in the left chest wall near the armpit with the size of 80 mm × 65 mm and the other was about 20 mm × 15 mm in the left supraclavicular. The qualitative were hard, with significant pain and poor activity. Local skin temperature was normal and not burst. No abnormal features were found during the physical examination of the lungs, heart, skin and other body systems.

Laboratory investigations revealed that complete blood counts, liver and renal function tests, and fasting blood glucose level and electrolyte panel were within normal limits. Needle biopsy showed that they were both metastatic adenocarcinoma (Fig. 1), chest CT revealed two low density occupied in left chest wall and left supraclavicular, they density were uneven, with areas of sac and irregular enhancement (Fig. 2). Ultrasound B revealed two heterogeneous low echoes which was irregular, blood flow could be seen in the signal and not oppression axillary vein (Fig. 3). The serum level of tumor markers (CEA,

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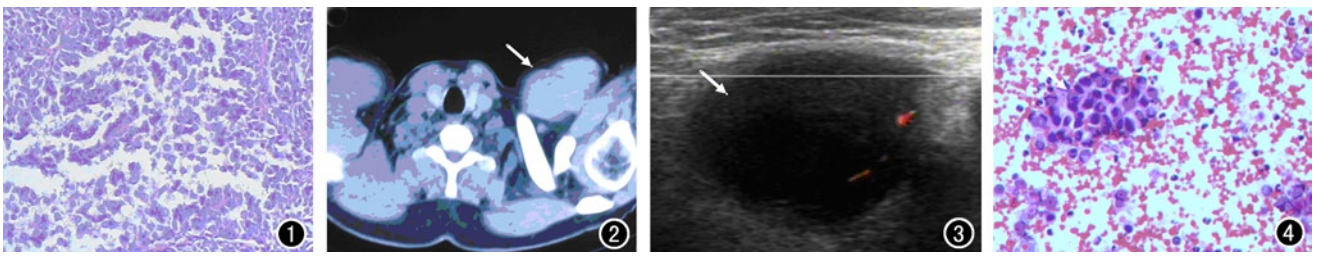

Fig. 1 Needle biopsy tips for metastatic adenocarcinoma (HE staining × 100)
Fig. 2 Chest CT revealed low density placeholders in the left chest wall near the armpit and left supraclavicular, their density was uneven, with areas of sac, strengthen the irregular
Fig. 3 Ultrasound B presented two heterogeneous low echoes in the left chest wall near the armpit and left supraclavicular. Shape is irregular, about 70 mm × 45 mm and 20 mm × 15 mm scope, blood flow can be seen in the signal, not oppression axillary and vein
Fig. 4 Histopathologic examination of the mass shows that the tumors' local was in sight of chrysanthemum form structure (HE staining × 100)

AFP) was in normal range and no obvious abnormal under nasopharyngeal, gastrointestinal and lung scrutiny.

In surgery, two gray red tumors located in the chest wall near the armpit and left supraclavicular which were invasion to the adjacent tissue. The edge was obvious edema and the tumor had rich blood supply. The section of the masses was a reddish gray zone and the cut was very brittle.

Histopathologic examination revealed that the tumor consisted of consistent small round cells, which presented a diffuse or papillary arrangement. Its local was in sight of chrysanthemum form structure (Fig. 4), and boundaries of cells was not clear. The nucleus were round, nucleolus were visible. Between cells it had more fiber vascular network and tumor cells were seen around the fiber adipose tissue.

Immunohistochemistry (IHC) staining showed that the tumor cells stained positively for CD99, neuron-specific enolase (NES), synaptophysin (Syn), S-100, and vimentin (VIM), suggestive of neuronal differentiation of the tumor. Tumor cells also expressed FLI-1, consistent with the diagnosis of pPNET. Other markers such as leukocyte common antigen (LCA), cytokeratin (CK) and Villin were negative. Fluorescence in situ hybridization and chromosomal study were not performed.

Discussion

In the beginning, pPNETs were not in the consideration, because of the result of needle biopsy was metastatic adenocarcinoma and this patient also had tinnitus, dizziness, diplopia, morning epostaxis, left supraclavicular node enlargrment. After taken nasopharyngoscope, chest CT, gastrointestinal barium series and some tumor markings, we excluded the possibility of nasopharyngeal carcinoma, lung cancer, mediastinal tumor and gastrointestinal tumor. We decided to perform radical resection to clear the masses pathology. Histopathologic examination told us the masses were pPNET, so we gave the patient symptomatic treatment. To review the clinical process and relevant literature, we presented some viewpoint as follows.

pPNETs is a rare disease which can be found in all age which usually can be seen before 35 years old and 20 in average [3]. It can appear in soft tissue, bone, retroperit, pelvic cavity, chest wall, lungs *et al.* It is highly biological invasive, recurrence or early metastasis which more transfer to the lungs, liver, kidney, even regional lympy node [4]. The mass general take on the shape of lobulated nodules, without a capsule and invasive growth. Its typical clinical manifestations is the mass grown quickly with pain and tumor-mass effects. Part can be associated with fever. In the present case, the growing quickly enormous masses caused pain and tumor-mass effects which had already impacted on raising of his left arm.

Image plays an important role in the diagnosis of patients with pPNETs, in which we can see the irregular soft tissue mass that is along the nerves. The mass is invasive strong. It grows in intramuscular spaces and packets of vesseles and nerves, which also can send more and cause tumor-mass effects. On chest CT, the mass is equal or low density. Its inner density is uneven when the mass is a bigger one, or high when its inner is bloody.There is aways no calcification inside, even if it has, it is often tiny [5]. And after enhancement, the calcification present different degree of reinforcement. To do Ultrasound B before operation is not only to realize the measurement of the mass, but also to make sure its inside blood supply and the positional relationship between the mass and the surrounding vasculature. Imaging feature of the present case presented similar results. Chest CT revealed low density placeholders in left chest wall and left supraclavicular, their density are uneven, with areas of sac, strengthen the irregular. Ultrasound B presented two heterogeneous low echoes can be seen in the left side of the chest wall near the armpit and left supraclavicular. Shape is irregu-

lar, about 70 mm × 45 mm and 20 mm × 15 mm scope, blood flow can be seen in the signal, not oppression axillary and vein.

The morphology characteristic was first described by Stout in 1918, and until 1993 it was determined to pPNETs by WHO [6]. Its histopathologic feature is that the cells of tumors are small and marked atypia with big and trachychromatic nucleus in which mitosis is obvious. The cells are round, oval shape or fusiform with less cytoplasm, in which can't find any glycogen. The special stain of PAS is negative. The tumor cells are solid and arranged in schistose, lobular, alveolar, linear. Tumors' local cells round blood vessels form in sight of chrysanthemum form structure without basal lamina. The centre of it is continuous filament, no cavity, no vessel, no sphacelus of tumor cells, and that is Homer-wright. Some scholars think [7] that Homer-wright is a significant evidence and characterisitic performance to diagnose pPNETs, of which two kind of neural markers are positive at least. It is positive to multiple markers such as CD99, NSE, Syn, S-100, Vimentin and negative to CK. Among these markers, CD99 has higher specificity which can be used to identify pPNETs and other small round cell tumors [8]. Recent research shows that Checking FLI-1 has higher application value for pPNETs [9]. In the present case, obvious Homer-wright had been seen. Its consequence of IHC was same as those reported in literature.

For most, the prognosis is poor, only 45% survive for five years, and 70% to 77% died. For pPNETs, research has shown that age is not the major factor [10] and measurement is not a underestimated factor influence prognosis. Prognosis is optimistic when patients are younger than 4, tumor`s location is in all fours or trunk, tumor`s measurement is less than 200 mm [11]. Currently there are no definite guidelines [12]. Presently, Operation treatment is still the first choice, but the simple operationexcision treatment effect is not ideal, one of the reason is that the tumor has already transferred before patients seeking medical advice. Another is the tumor is most located in trunk which is difficult to implement complete resection. Saeed [13] think mass removal followed by adjuvant chemo-radiation is associated with the best clinical consequence. Therefore, most patients choose the comprehensive treatment with Radical resection in combination with local radiotherapy and chemotherapy [14]. Duan [15] think autologous stem cell rescue besides adjuvant chemotherapy has been associated with prolonged survival. For the present case, because of the preoperative needle biopsy confused diagnosis, we had not given him adjuvant chemo-radiation. After getting rid of possible primary organs and making sure that the mass can be complete resection, the tumor perform radical resection. Pathologyc reveals the diagnosis of pPNETs in the postoperative, which could receive chemotherapy by Cyclophosphamide, Doxorubicin, Vincristine and high-does Cisplatin and local radiotherapy. Now one year after operation, the living conditions of patients with good.

Conflicts of interest

The authors indicated no potential conflicts of interest.

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