Case report - Thoracic oncologic

Primitive chest wall neuroectodermal tumor in a pediatric patient

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Abstract

A 13-year-old boy with a primitive neuroectodermal tumor of the chest wall is presented. After four cycles of chemotherapy, a computed tomography scan of his chest showed a larger mass invading the left upper lobe of the lung. He underwent resection of the left chest wall from the left fourth to sixth ribs, including the tumor, combined with left upper lobectomy and lymph node dissection. A diagnosis of primitive neuroectodermal tumor was confirmed histopathologically and immunohistochemically. After surgery, four cycles of chemotherapy with ifosfamide and etoposide were given. One year after treatment, the patient is currently doing well without evidence of recurrence.

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1. Introduction

Primitive neuroectodermal tumor (PNET) of chest wall origin is a rare malignant tumor in children and young adults [1]. The mainstay of treatment for PNET is doxorubicin-based chemotherapy; however, surgical resection is still necessary for a definite diagnosis and to avoid tumor invasion of the vital organs. Moreover, complete resection with negative margins followed by chemotherapy may achieve a longer tumor-free survival time [2]. A case of PNET of the chest wall invading the lung with regional lymph node metastasis, which was treated by initial chemotherapy, resection and adjuvant chemotherapy, is described.

2. Case report

A 13-year-old boy presented with a left chest mass about 8.0 cm in maximum diameter with tenderness. A chest X-ray and computed tomography (CT) scan revealed an abnormal shadow on his left chest wall. Small, round malignant cells were identified following needle biopsy, but a definite diagnosis could not be made because of the small sample size. From the clinical presentation and cytological features, PNET was suspected.

A sound clinical response was achieved after two cycles of chemotherapy with ifosfamide, vincristine and doxorubicin. However, after the fourth cycle, a chest CT-scan showed a larger mass on the left chest wall invading the left upper lobe (Fig. 1a,b). Positron emission CT showed increased fluorodeoxyglucose uptake within the mass (a standardized uptake value maximum of 5.3; Fig. 1c) with no evidence of distant metastasis. Because of the patient’s good performance status, the size and location of the tumor, and the inefficacy of the initial chemotherapy, it was decided to proceed with surgical resection.

En bloc resection of 10 cm×20 cm of the chest wall from the left fourth to sixth rib, including the tumor, combined with left upper lobectomy was performed, along with mediastinal and hilar lymph node dissection. The chest wall was reconstructed using expanded polytetrafluoroethylene sheet.

Histopathologically, small round cell proliferation was observed in masses from the chest wall, left upper lobe and pulmonary ligament lymph node; no tumor cells were identified in the resected margins. Immunohistochemical staining showed that the tumor was expressing CD99, MIC2, vimentin, neurofilament, synaptophysin, CD10, CDR45 and Ki-67, but was negative for the expression of pancytokeratin, cytokeratin 7, cytokeratin 20, chromogranin, CD3, CD20, CD30, CD56, desmin, S-100, and prostate-specific antigen. In line with these findings, a diagnosis of PNET was confirmed.

The patient received four cycles of postoperative combination chemotherapy consisting of ifosfamide and etoposide. One year after treatment, the patient is currently well doing without evidence of recurrence.

3. Discussion

PNETs, part of the Ewing’s sarcoma family of tumors [3], are malignant tumors of small undifferentiated neuroectodermal cells thought to originate from neural crest cells, and may develop in soft tissue. The diagnosis of PNET is
based on the presence of small round cells with neural differentiation [1]; however, this may be difficult to achieve due to the small sample size obtainable [4]. Cytogenetic analysis has become the standard for confirming the diagnosis. Most Ewing’s sarcomas have a stereotypical immunophenotype, with the expression of CD99 in all cases and no expression of cytokeratins or desmin [5].

The mainstay of treatment for PNET is doxorubicin-based chemotherapy. For non-metastatic PNET, overall survival is improved with multimodality therapy consisting of chemotherapy, resection and radiotherapy. Nevertheless, when initial chemotherapy is not efficacious and successful resection with negative tumor margins is anticipated, resection followed by chemotherapy for PNET of chest wall origin may be the treatment strategy of choice, both for confirming the diagnosis and for preventing invasion to other tissues that might be fatal [6].

The decision of surgical resection in our case was based on the preoperative clinical and radiographic presence of tumor, which, by defining a larger mass in the chest wall invading the left upper lobe of the lung, showed initial chemotherapy to have been ineffectual. Definitive diagnosis was made histopathologically and immunohistochemically. As PNET is an aggressive tumor, metastases to lymph nodes within the bronchial tree should be considered, and a positive lymph node consequently changed the stage. The patient received combination chemotherapy and has experienced no recurrence for one year. Complete resection with intensive chemotherapy thus achieved a cure of chest wall PNET with accompanying lymph node metastasis.

4. Conclusion

In conclusion, when resection with negative tumor margins is anticipated, surgical resection could be considered in cases of PNET with unsatisfied initial chemotherapy.

References