Primitive Neuroectodermal Tumor of the Breast: About 2 Cases

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Introduction

Neuroendocrine (NE) breast cancers encompass a heterogeneous group of tumours showing morphological features similar to those of NE neoplasms of the gut and lung and expressing one or more neuroendocrine markers (neuron specific enolase, chromogranins synaptophysin) in at least 50% of tumour cells. They are rare lesions representing about 2–3% of all breast cancers. The coexistence of both exocrine and neuroendocrine differentiation can lead to some uncertainty about the choice of the best antineoplastic strategy.

Case1: A 59-year-old woman was admitted to hospital with a tumor on her right breast. Mammography and ultrasonography reported a lesion measuring 3 cm. CT scan of the chest and abdomen found no abnormalities.Histopathological examination of the specimen obtained from a biopsy of the lesion detected an invasive breast ductal carcinoma. The patient underwent right mamectomy with axillary lymph node dissection. The final histopathologic diagnosis was primitive neuroectodermal tumor with 1N+/20N.Immunohistochemical detected synap tophysin(+),andcytokeratin(+),chromograninA(-) ,and PR(+) ER(-),Her2/neu (score1+). Adjuvant treatment consisted of chemotherapy,and radiotherapy to the chest wall. And additional hormonotherapy.The patient was followed-up every 3 months. Two years after treatment no recurrence was detected.

Case2: A 37year old woman presented with a tumor in her right breast. Mammography and ultrasonography reported a lesion measuring 3 cm.Needle aspiration biopsy detected carcinoma cells. No distant metastasis was detected on various investigations such as chest XRay,liver ultrasond,bone scan and CT chest. The patient underwent tumorectomy with axillary lymph node dissection. The final histopathologic diagnosis was primitive neuroectodermal tumor with 20N-/20N. Immunohistochemical detected cytokeratin(-),chromograninA(+) ,NSE(+)and PR (-) ER (-). Her2/neu (-). Adjuvant treatment consisted of chemotherapy,and radiotherapy. Two years after treatment no recurrence was detected.

Discussion

Neuroendocrine tumors in the breast are rare, accounting for less than 0.1% of all breast cancers and less than 1% of all neuroendocrine tumors.Neuroendocrine carcinomas of the breast occurred mainly in older women around the end of the seventh decade of life. The clinical and radiological features of neuroendocrine tumors are non specific, and fine-needle aspiration or core-needle biopsy examination is necessary for the diagnosis.The morphological features of the neuroendocrine carcinomas of the breast should be confirmed by immunohistochemical means or by electron microscopy. Immunohistochemically, NSE was used to study argyrophilic breast carcinoma, and it has been found to be positive in 16–50% of unselected breast tumors, even in the absence of argyrophilia or dense core granules.All these findings discouraged us from using NSE to define a breast carcinoma as neuroendocrine differentiated. In contrast to NSE, chromogranin and synaptophysin have been widely accepted as specific markers of neuroendocrine differentiation. Neuroendocrine tumor in the breast may represent either metastatic or primary lesions. The ductal carcinoma in situ component is the only absolute proof of the primary nature of the breast carcinoma. It is important to differentiate primary breast neuroendocrine tumor from metastatic disease to the breast because of differences in treatment.

Treatment, which may include surgery, radiotherapy, and chemotherapy, is based on clinical stage and the presence of metastases. Prognosis is variable and is dependent on the initial stage of disease.

Conclusion

Primary neuroendocrine carcinoma of the breast is rare-only about 30 cases have been reported in literature.and it is important that these be recognized as a separate entity from the more common breast carcinoma keeping in mind the difference in behavior of the two tumors for planning therapy.