Rhinology, 38, 144–146, 2000

Olfactory neuroblastoma metastatic to the breast*

Subhash Chandra Shetty[1], Sanjeev Gupta[1], Geetha Chary[1], Shameem Shariff[2], Vinutha Acharya[2], Belliappa M.S.[3]

[1] Department of Otorhinolaryngology-Head and Neck Surgery, St Johns Medical College Hospital, Bangalore, India
[2] Department of Pathology, St Johns Medical College Hospital, Bangalore, India
[3] Curie Centre of Oncology, St Johns Medical College Hospital, Bangalore, India

SUMMARY

Olfactory neuroblastoma (Esthesioneuroblastoma) is a rare malignant tumour arising from olfactory epithelium. It has a predilection for cervical lymph node metastasis and also has potential for distant metastasis to unusual sites like scalp, face, aorta, spleen, liver, adrenal gland and ovary. We report here a rare case of olfactory neuroblastoma in an adolescent girl with metastatic deposits in the breast. A poor prognosis due to rapidly progressive disseminated disease was observed. The relevant literature regarding metastatic olfactory neuroblastoma and metastasis in the breast from non-mammary malignant neoplasms is reviewed.

Key words: olfactory neuroblastoma, metastasis, breast

INTRODUCTION

Olfactory neuroblastoma (ONB) is a rare malignant tumour of the nasal cavity and anterior skull base that arises from the basal cells of the olfactory neuroepithelium. ONB is a locally aggressive tumour with frequent local recurrences, metastasis occurs in about 10 to 30% of the patients (Skolnick et al., 1966). The most commonly involved sites are the cervical lymph nodes (Lewis et al., 1965) but the lungs (Mendeloff, 1957), long bones (Hutter, 1963) and pelvis are the other reported sites. There are isolated reports of metastasis to the scalp (Walters et al., 1980), orbit (Grahne, 1965), aorta, spleen, liver, adrenal gland and ovary (Wade et al., 1984). We report here an unusual case of ONB with metastasis to the breast in a young woman.

CASE REPORT

A 13-year old female presented to us with left facial pain, nasal obstruction, epiphora, anosmia and recurrent epistaxis of two weeks duration. Physical examination revealed a polypoidal, fleshy nasal mass and swelling in the left maxillary and infraorbital region. Examination of the neck revealed ipsilateral palpable lymph nodes. A contrast enhanced computerised tomography revealed an extensive enhancing lesion in the anterior cranial fossa invading the nasal cavity, ethmoid sinus, left sphenoid sinus, invading the cribriform plate and elevating the dura under the left frontal lobe. Nasal biopsy and Fine needle aspiration cytology (FNAC) of cervical lymph nodes were performed and histopathological examination of the biopsied nasal mass showed a cellular neoplasm composed of cells slightly larger than lymphocytes with the nuclei showing a fine granular chromatin. The cells were arranged in clusters, separated by fibrous stroma (Figure 1). Some of the cells showed fibrils extending from the cytoplasm. FNAC of cervical lymph node was consistent with ONB. She was planned for 55Gy of external beam radiotherapy over a six-week period followed by chemotherapy with Cyclophosphamide and Vincristine. During the course of radiotherapy she developed swellings in the right breast, left axilla and back. FNAC of the breast and axilla showed metasta-

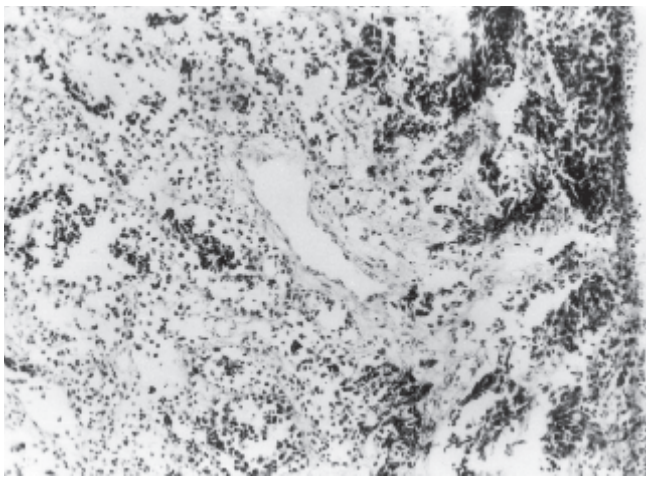

Figure 1. Photomicrograph showing neoplasm with small round cells arranged in clusters and seemingly lining spaces separated by fibrous stroma. Fibrillary character of the cytoplasm was observed. (Haematoxylin and Eosin ×25)

* Received for publication September 27, 1999; accepted March 10, 2000

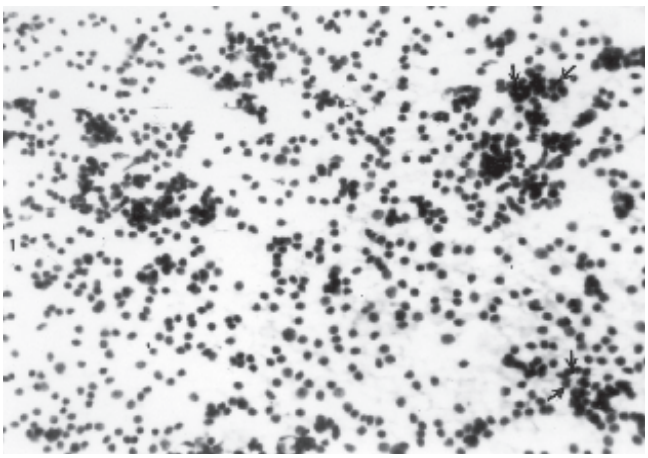

Figure 2. Smear shows a cellular spread composed of sheets of circular cells with nuclear pleomorphism. An attempt at Horner Wright rosette formation is observed (arrows). (Haematoxylin and Eosin ×250)

tic ONB with the picture of round cells with dark staining nuclei dispersed among red blood cells. Some of the cells were arranged in ill-formed rosettes with a few neurofibrillary tangles (Figure 2). She also developed bone marrow metastasis and multiple deposits in the right ovary in the same period. The patient died of disseminated disease subsequently.

DISCUSSION

ONB is a rare tumour in the nasal cavity and appears at all ages from childhood to elderly people. Age incidence has bimodal distribution with peaks at 11-20 years and 40-60 years (Elkon et al., 1979). This tumour was originally reported by Berger et al., in 1924. The most widely utilised staging system was developed by Kadish et al., in 1976. The stages are: stage A - involvement of the nasal cavity only; stage B - involvement of the nasal cavity plus one or more paranasal sinuses; stage C - involvement beyond the nasal cavity, including involvement of the orbit, base of skull or intracranial cavity, cervical nodes or distant metastatic sites.

The tumour is usually slow growing, developing local recurrence very frequently (50%). Distant metastasis has reportedly occurred in about 16 % (Bolla et al., 1980) to 33% (Olsen and DeSanto 1983) of cases. The metastatic disease and locally aggressive behaviour of ONB worsens its prognosis and groups it under Group C of the Kadish staging system. Performance of fine needle aspiration cytology of all suspicious masses in the cervical region and other sites prior to the institution of therapy is of importance in evaluation of distant metastasis.

Immunohistochemistry is important in separating the ONB from other similar sinonasal neoplasms like undifferentiated carcinoma, lymphoma, melanoma, or sarcoma. Neoplastic cells exhibit positive immunohistochemical staining reactions with antibodies to neuron-specific enolase neurofilaments, class III beta-tubulin isotype, microtubule-associated protein, and synaptophysin. Some cases show a distinctive peripheral staining pattern with antibodies to the S-100 protein. There is nonetheless a considerable variation in staining (Morita et al., 1993). The diagnosis of metastatic lesion can also be clarified by immunohistochemical demonstration of the unique antigenic profile that can be obtained in routine processed biopsies.

Most of the authors state that surgery is the first choice of treatment followed by radiation or combined therapy when recurrences occur (Skolnick et al., 1966; Elkon et al., 1979). A combined craniofacial resection is the surgery of choice in patients with no metastasis. Craniofacial resection in conjunction with radical neck dissection can be performed in unilateral cervical metastasis (Levine et al., 1986). Absolute contraindications for aggressive surgical therapy include diffuse metastatic disease and uncontrolled second primary tumour. Chemotherapy with cyclophosphamide and vincristine should be reserved for the most advanced cases (Akira et al., 1984).

Prognosis of this tumour has been considered to be good i.e., for the tumour confined to the nasal cavity, 3 year survival rate is 100%; and the least, about 40% for a tumour with distant metastasis (Kadish et al., 1976).

Metastasis to the breast from non-mammary malignant neoplasms are rare, accounting for approximately 0.5% to 1.4% of breast tumours (Toombs and Kalisher, 1977). Malignant melanoma and lung carcinoma are the most commonly reported primary tumours metastatic to the breast, while neoplasms of the ovary, stomach, kidney, prostate, oropharynx, and other sites are less frequent sources of metastasis (Hajdu and Urban 1972; Toombs and Kalisher, 1977). In children, rhabdomyosarcoma is the most common (Howarth et al., 1980). Boothroyd reported a solitary case of metastatic neuroblastoma in a study of 17 cases of breast masses in childhood and adolescence over a five-year period (Boothroyd and Carty, 1994). Metastases in the breast are usually painless upper outer quadrant masses. On mammography they are typically well-circumscribed lesions without microcalcifications (Bohman et al., 1982). A breast mass in a patient with a history of cancer, even if clinically or mammographically benign, should raise suspicion of a metastasis. Pathological features include a pattern similar to the primary neoplasm and an absence of in situ carcinoma, which characterises primary breast cancer (Amichetti et al., 1990). Accurate diagnosis of the breast metastasis is important to avoid unnecessary mastectomy and to implement an appropriate systemic therapy. Metastases in the breast almost always indicate advanced systemic disease and have an extremely poor prognosis as in the present case.

CONCLUSION

Olfactory neuroblastoma is a rare tumour arising from the olfactory neuroepithelium with high incidence of local recurrence. Distant metastasis occurs most commonly to the cervical lymph nodes, lung and a few other rare sites. Our case report highlights the unusual site of metastasis from olfactory neuroblastoma to the breast in an adolescent girl.

REFERENCES

1. Akira T, Makimoto K, Okawa M, Hirono Y, Yamabe H (1984) Olfactory neuroblastoma; Presentation of a case and review of the Japanese literature. Laryngoscope 94:252-256.
2. Amichetti M, Perani B, Boi S (1990) Metastases to the breast from extramammary malignancies. Oncology 47:257-260.

3. Berger L, Luc, Richard (1924) Lesthesio-neuroepitheliome olfactif. Bulletin Assoc. Franc. Et. Cancer 13:410-421.
4. Bohman LG, Basset W, Gold RH, Voet R (1982) Breast metastases from extramammary malignancies. Radiology 144:309-312.
5. Bolla M, Vroussos C, Roux O (1980) Les Esthesioneuromes Olfactifs. Revue Generale a Propos de Trois Nouvelles Observations. Bull. Cancer, (Paris) 67:325-332.
6. Boothroyd A, Carty H (1994) Breast masses in childhood & adolescence. A presentation of 17 cases and a review of literature. Paediatric Radiology 24:81-84.
7. Elkon D, Hightower SI, Lim ML (1979) Esthesioneuroblastoma. Cancer 44:1087-1094.
8. Grahne B (1965) Olfactory neuroblastoma. Acta Otolaryngology 59:55-60.
9. Hajdu SI, Urban JA. (1972) Cancer Metastatic to the Breast. Cancer 29:1691-1696.
10. Howarth CB, Caces JN, Pratt CH (1980) Breast metastases in children with rhabdomyosarcoma. Cancer 46:2520-2524.
11. Hutter RVP (1963) Esthesioneuroblastoma: A clinical and pathological study. American Journal of Surgery 106:748-753.
12. Kadish S, Goodman M, Wang CC (1976) Olfactory neuroblastoma: a clinical analysis of 17 cases. Cancer 37:1571-1576.
13. Levine PA, Mclean WC, Cantrell RW (1986) Esthesioneuroblastoma: The University of Virginia experience 1960-1985. Laryngoscope 96:742-746.
14. Lewis JS, Hutter RVP, Tollefsen HR (1965) Nasal tumours of olfactory origin. Archives of Otolaryngology 81:169-174.
15. Mendeloff J (1957) The oflactory neuroepithelial tumours. Cancer 10:944-956.
16. Morita A, Ebersold MJ, Olsen KD, Foote RL, Lewis JE, Lynn M, Quast RN (1993) Esthesioneuroblastoma: Prognosis and Management. Neurosurgery 32(5):706-714.
17. Olsen KD, DeSanto LW (1983) Olfactory Neuroblastoma. Archives of Otolaryngology; 109:797-802.
18. Skolnick EM, Massari FS, Tenta LT (1966) Olfactory Neuroepithelioma. Review of the World Literature and presentation of two cases. Archives of Otolaryngology 84:644-653.
19. Toombs BD, Kalisher L (1977) Metastatic disease to the breast. Clinical, pathologic and radiographic features. American Journal of Roentogenology 129:673-676.
20. Wade PM, Smith RE, Johns ME (1984) Response of Esthesioneuroblastoma to Chemotherapy; Report of Five cases and Review of the literature. Cancer 53:1036-1041.
21. Walters TR, Puspharaj N, Ghander AZ (1980) Olfactory neuroblastoma. Archives of Otolaryngology 106:242-243.

Dr. Subhash Chandra Shetty
Department of Otolaryngology and Head and Neck Surgery,
St Johns Medical College Hospital,
Bangalore- 560 034. India
Fax: 91-80-5530070.
E-mail: stjohns@blr.vsnl.net.in