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PATHOLOGIC QUIZ CASE 1

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A 69-year-old man complained of intermittent left-sided epistaxis and nasal obstruction for several years. Physical examination disclosed a

pink-gray polypoid mass filling the upper left nasal cavity. The right nasal cavity and nasopharynx were clear. Multiple biopsy specimens were

obtained. The microscopic findings are shown in Figs 1 and 2.

What is your diagnosis?

PATHOLOGIC QUIZ CASE 1

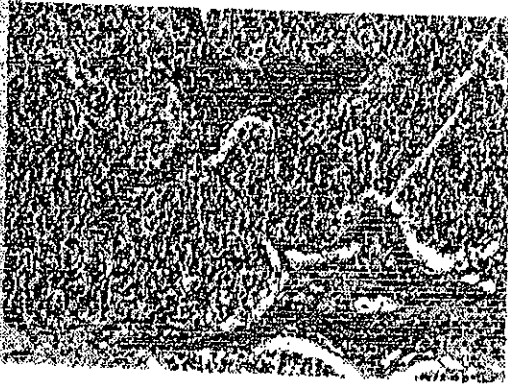


Figure 1.

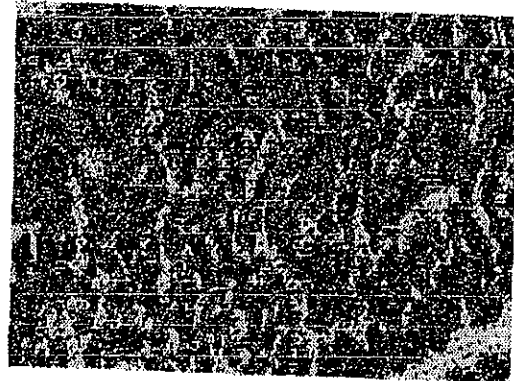


Figure 2.

Microscopic findings
in Figs. 1
and 2.
Diagnosis?

PATHOLOGIC QUIZ CASE 2

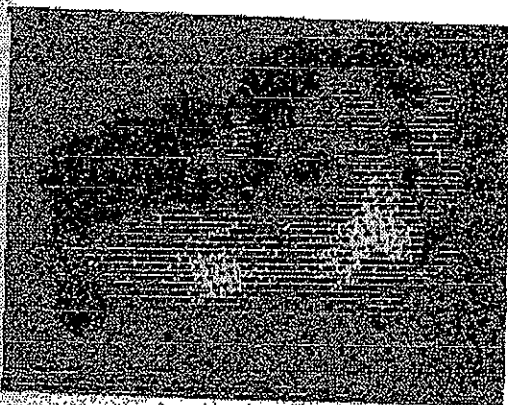


Figure 1.

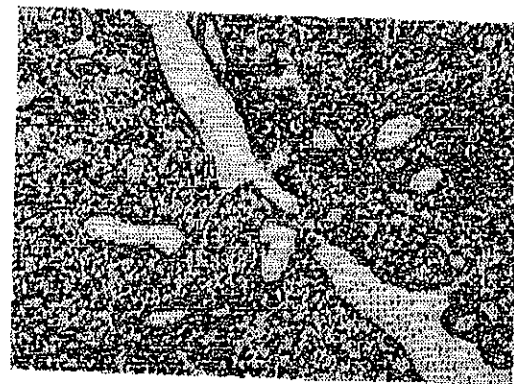


Figure 2.

A mass in the nasopharynx
was biopsied, nonulcerated,
and removed from the right middle
turbinate (Fig. 1) was surgically
removed. The specimen
was submitted for
histopathologic exami-
nation (Figs. 2 and 3).
Diagnosis?



Figure 3.

The
patient
was
operated
on
for
the
nasopharyngeal
carcinoma.

PATHOLOGIC QUIZ CASE I

Pathologic Diagnosis: Olfactory neuroblastoma.

The diagnosis of olfactory neuroblastoma can usually be made by light microscopy. The presence of a fibrillary intracellular background is needed for a conclusive diagnosis. The fibrils have been shown ultrastructurally to represent neuronal cell processes.¹

In the undifferentiated olfactory neuroblastoma, electron microscopy is often needed to distinguish it from embryonal rhabdomyosarcoma, undifferentiated lymphomas, undifferentiated epidermoid carcinoma, plasmacytomas, transitional cell carcinoma, and others. Electron microscopy can detect neurites and secretory granules compatible with catecholamine granules that are diagnostic of olfactory neuroblastoma.² Sympathetic neuroblastoma and a typical olfactory neuroblastoma are virtually identical by light microscopy. The similarity continues at the ultrastructural level, indicating that the ultrastructural characteristics of neuroblastoma are similar regardless of location or presumed origin.³ Micheau and coworkers⁴ demonstrated the presence of dopamine- β -hydroxylase and catecholamines, using immunohistochemical and biochemical studies, indicating the likelihood that olfactory neuroblastoma is of sympathetic origin. They recommended assays of urinary vanillylmandelic acid (VMA), dopamine, and homovanillic acid to gain further information about the nature of olfactory neuroblastoma and its diagnosis. Taxy and Hidvegi⁵ noted that the level of β -hydroxylase in olfactory neuroblastoma is 100 times less than that observed in sympathetic neuroblastoma. No systemic manifestations of catecholamines from olfactory neuroblastoma have been noted.

Although it is a slow-growing tumor, olfactory neuroblastoma has a

significant rate of metastasis. Bailey and Barton⁶ noted a 24% incidence (40/165 patients) of metastases. The most common sites, in order of decreasing frequency, are the cervical lymph nodes, lungs and pleura, long bones, spinal column, breast, and abdominal viscera. Death results from distant or local metastasis or intracranial extension leading to meningitis and/or hemorrhage.

Treatment for olfactory neuroblastoma has remained controversial. In a review involving 97 cases, Skolnik et al⁷ reported five-year survival rates of 64% with radical surgery, 38% with radiation, and 50% using a combination of surgery and radiation. They recommend surgery, reserving radiation for persistent or recurrent tumors or unresectable lesions. Cantrell et al⁸ recommended combined therapy, including preoperative radiation and radical surgery.

Doyle and Paxton⁹ used a combined neurosurgical and otolaryngologic approach to olfactory neuroblastoma. A craniotomy performed by neurosurgery would enable assessment of the resectability of the tumor. The procedure could then be abandoned if indicated. If appropriate, an en-bloc excision of the cribriform plate and pansinusectomy could be performed.¹ The defect is repaired using preserved dural graft. They also advised radical neck dissection in cases with clinical cervical adenopathy.

The surgical management as described by Bailey and Barton⁶ emphasized adequate excision of tumor, usually through a lateral rhinotomy approach with resection of the lateral wall of the nose, including tumor extending into the ethmoid cells, sphenoid, orbit, or pterygoid space and, in some cases, resection of the cribriform plate. Chemotherapy has not been sufficiently used to properly assess its efficacy in treating olfacto-

ry neuroblastoma.

Olfactory neuroblastoma is a rare malignancy of the head and neck that often requires diagnosis through the use of electron microscopy. The work-up must involve accurate radiographic imaging to assess the extent of the tumor. Particular attention must be given to whether or not the cribriform plate is involved. If there is involvement of the cribriform plate, then a combined otolaryngologic-neurosurgical procedure should be used, with en-bloc resection of the cribriform plate combined with removal of the appropriate paranasal sinuses and wall of the nose. If the cribriform plate is not involved, then the preferred surgical approach is through a lateral rhinotomy. Furthermore, if clinical cervical adenopathy is present, a radical neck dissection should be performed. With evidence of distant metastatic disease or in the case of an unresectable lesion, radiation should be used. Radiation may be used for recurrence as well. Lifelong follow-up examination is essential to rule out recurrence or persistence.

References

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