CT AND ANGIOGRAPHY OF ESTHESIONEUROBLASTOMA

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ABSTRACT

A case report of Esthesioneuroblastoma in a 39-year-old female patient was presented. The mass was a slow growing one, when found it was quite extensive to be located in both nasal cavities, medial part of right maxillary sinus, both ethmoid sinuses, left sphenoid sinus, left orbital cavity, and epidural space of the anterior cranial fossa. Bowing pattern, bony erosion and tumoral calcification was shown by CT scan. The metastatic tumor to right parotid gland was already present. The tumor received blood supply from both maxillary arteries and left ophthalmic artery and the main feeder was left maxillary artery which indicated that the tumor originated from the left side of the nasal cavity.

INTRODUCTION

Esthesioneuroblastoma is a rare nasal neoplasm, arising from neuroepithelial elements in the olfactory membrane in the superior portion of the nasal cavity (1). It occurs in all ages, with a range of 3 to 79 years of age. The incidence shows a bimodal distribution with peaks in the second and sixth decades of life (2). The distribution between the sexes is roughly equal. The symptoms are non-specific and include epistaxis, anosmia, rhinorrhea and nasal obstruction. Due to this lack of symptoms, most patients are diagnosed late in the course of the disease (1). It usually appears as a red or fleshy mass in the nasal vault. Symptoms of local invasion, such as proptosis or headache, are usually evident at diagnosis.

The tumor was first described as 'L' esthesioneuroepitheliome olfactif' in 1924 by Berger (3). Numerous names have been used to describe this neoplasm including olfactory neuroblastoma, olfactory esthesioneuroma, esthesioneurocytoma and neuroendocrine carcinoma (4).

We present a case of this tumor by CT and angiographic imaging.

CASE REPORT

A 39-year-old female patient from Pracheenburi province, was referred to Rajvithi hospital due to the presence of the intranasal mass. She had the symptom of nasal congestion for 10 years and the left intranasal mass was palpated later. Another mass was palpated at right cheek for 8 months. Axial and coronal CT scan of the nasal cavities was performed and showed a 5.5X7X5.3 cm enhanced soft tissue density mass with an epicenter in the nasal cavities. The growth of the tumor was seen in both nasal cavities, medial part of right maxillary sinus, subcutaneous fat plane and skin of the nose, in both ethmoid sinuses, anterior part of left sphenoid sinus and medial part of left orbital cavity. Expanding appearance of the mass was appreciated at medial walls of both maxillary sinuses, in right ethmoid sinus and medial wall of ethmoid sinus. Dense calcification was shown in the central part of the mass. Small extension to epidural space of anterior cranial fossa was noted. Another tumor mass was seen at the superficial portion of right parotid gland, size 3 X 3.5 X 4 cm with well defined border. The cavernous sinuses were normal. Local

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destruction of the hard palate was observed (Fig.1,2).

Biopsy from the mass in left nasal cavity revealed small round cell tumor. The study of immunohistochemical one led to the diagnosis of olfactory neuroblastoma by Dr. Arunluck Komindr, the pathologist of Rajvithi Hospital. Metastatic nodule at right parotid gland from esthesioneuroblastoma was also diagnosed from the biopsy by the same pathologist.

The patient was sent to Ramathibodi hospital for pre-surgical embolisation. Pre-embolized angiography revealed a hypervascular large nasal mass which received bloody supply from left maxillary artery (main feeder), right maxillary artery and left ophthalmic artery. Successful Ivalon and gelfoam embolization of both maxillary arteries was shown (Fig.3,4).

DISCUSSION

Because the normal distribution of olfactory epithelium may extend from the cribriform plate to the level of the middle turbinates, esthesioneuroblastoma may arise in the region of the nasal cavity anywhere throughout this distribution (4,5). Light microscopic study reveals features similar to classical childhood neuroblastoma. In an upper nasal neoplasm, the presence of a fibrillary intercellular background in conjunction with Homer-Wright pseudorosettes is considered to be diagnostic of olfactory neuroblastoma. However, these hallmarks are not always evident, and confusion with other primary tumors of the nasal cavity and paranasal sinuses, such as lymphoma, undifferen-tiated carcinoma, and extramedullary plasmacytoma is possible. Therefore, immunohisto-chemistry and electron microscopy are necessary for the histologic diagnosis (1).

Although slow growing, esthesioneuroblastomas are locally invasive and can metastasize to regional lymph nodes, lung or bone. No treatment had been convincingly (1,2,5,6). Survival is related to the stage of disease at initial diagnosis (2,4). Kadisch (6,7) proposed a staging classification of esthesioneuroblastoma based on extent of disease: stage A is involvement of the nasal cavity only; stage B is involvement of the nasal cavity and one or more paranasal sinus; and stage C is involvement outside the nasal cavity including orbit, base of the skull, intracranial cavity, cervical nodes, or distant metastases.

The CT features in 9 patients of this tumor studied by Hurst (4) demonstrate a fairly consistent pattern. All tumors were centered in the superior nasal cavity or ethmoids. Tumor density prior to enhancement was relatively homogeneous and was equal to or greater than the surrounding soft tissue. Contrast enhancement was usually moderate in intensity and homogeneous but, with one exception, was without cystic or hemorrhagic areas. Calcification has been reported. The location and size of the calcific densities often made it difficult to determine radiologically whether this represented tumoral calcification or bony fragments secondary to destruction by tumor. Bony erosion was often associated with bowing. Intracranial extension was seen in 20-57% and was very common is Stage C (4,8). All stage C cases demonstrated involvement of the orbit. Displacement of the medial rectus muscle by tumor mass was in present in 81%. The periorbital tissue is frequently acts as a barrier.

The MR features of the tumor are nonspecific and is variable (7). The tumor can have homogenous or heterogeneous signal characteristics. Compared with brain gray matter, the tumors are generally hypointense on T1WI and isointense to hyperintense on PD and T2WI. Contrast enhancement of the tumor is variable but always present. It often expands the nasal cavity, usually with concurrent destruction of the nasal septum turbinates, and ethmoid septa. Local extension with further bone destruction can occur.

Our patient should be classified as Stage C. The CT findings are similar to reported cases, including features of tumoral calcification, bony erosion and bowing pattern.

In the series of 26 patients, reported by Levine (9), an increased disease-free interval exists with the use of combined preoperative radiotherapy and craniofacial resection for stage A and B disease, and the addition of preoperative and postoperative vincristine and Cytoxan for stage C disease.

ACKNOWLEDGMENTS

We thank the department of Radiology of Rajvithi Hospital for the CT images.

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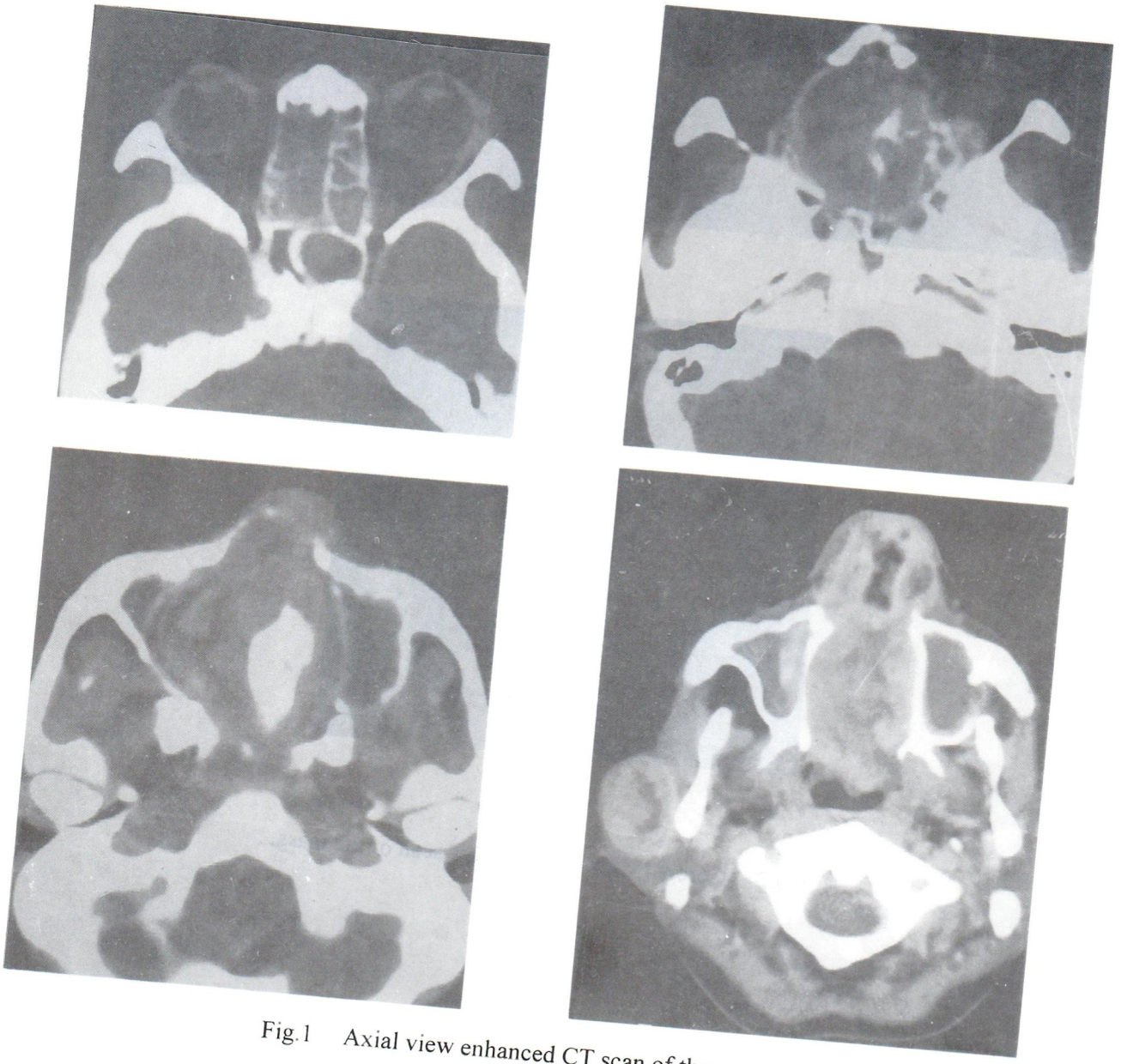

Fig.1 Axial view enhanced CT scan of the mass

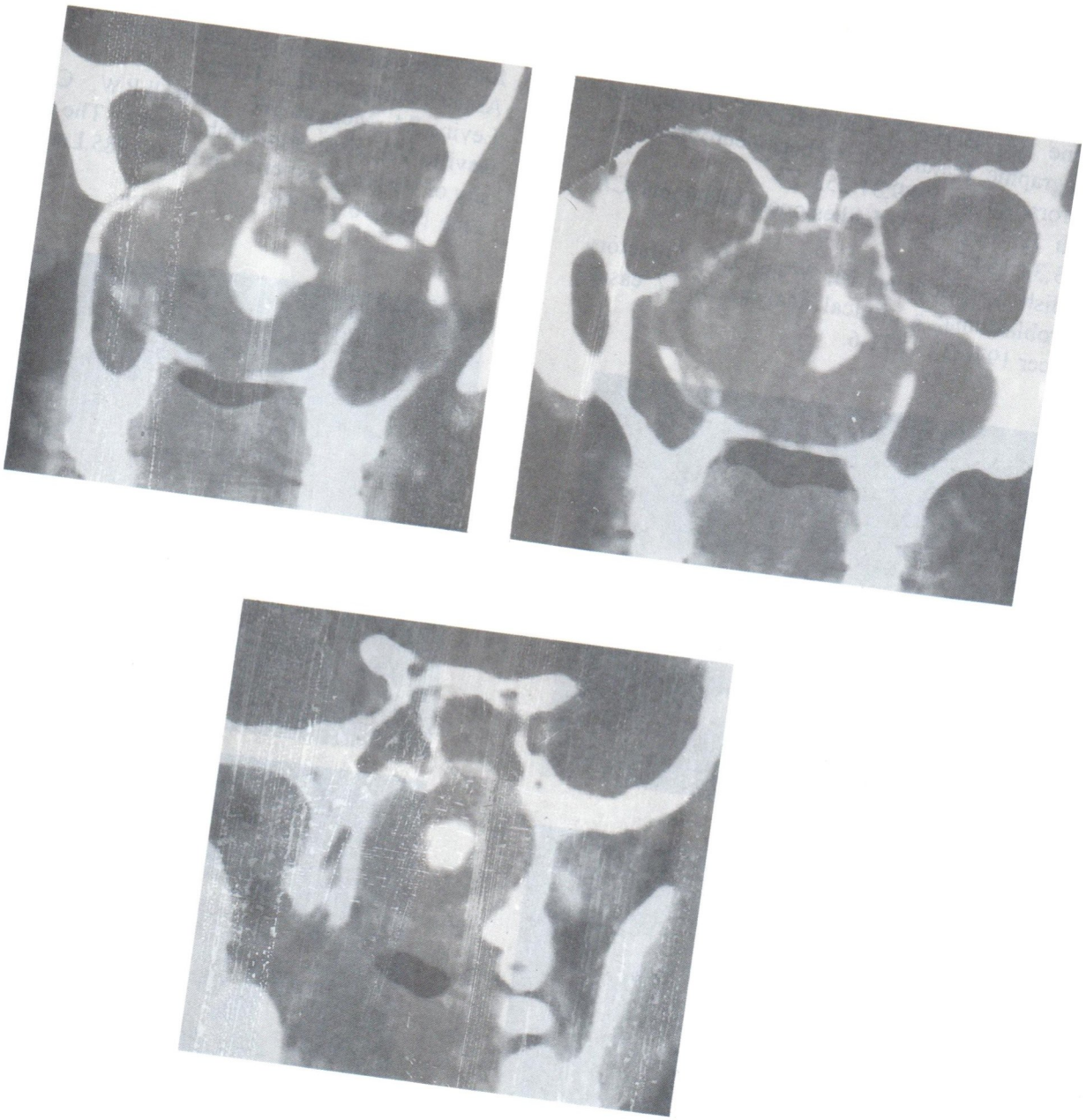

Fig.2 Coronal view enhanced CT scan of the mass

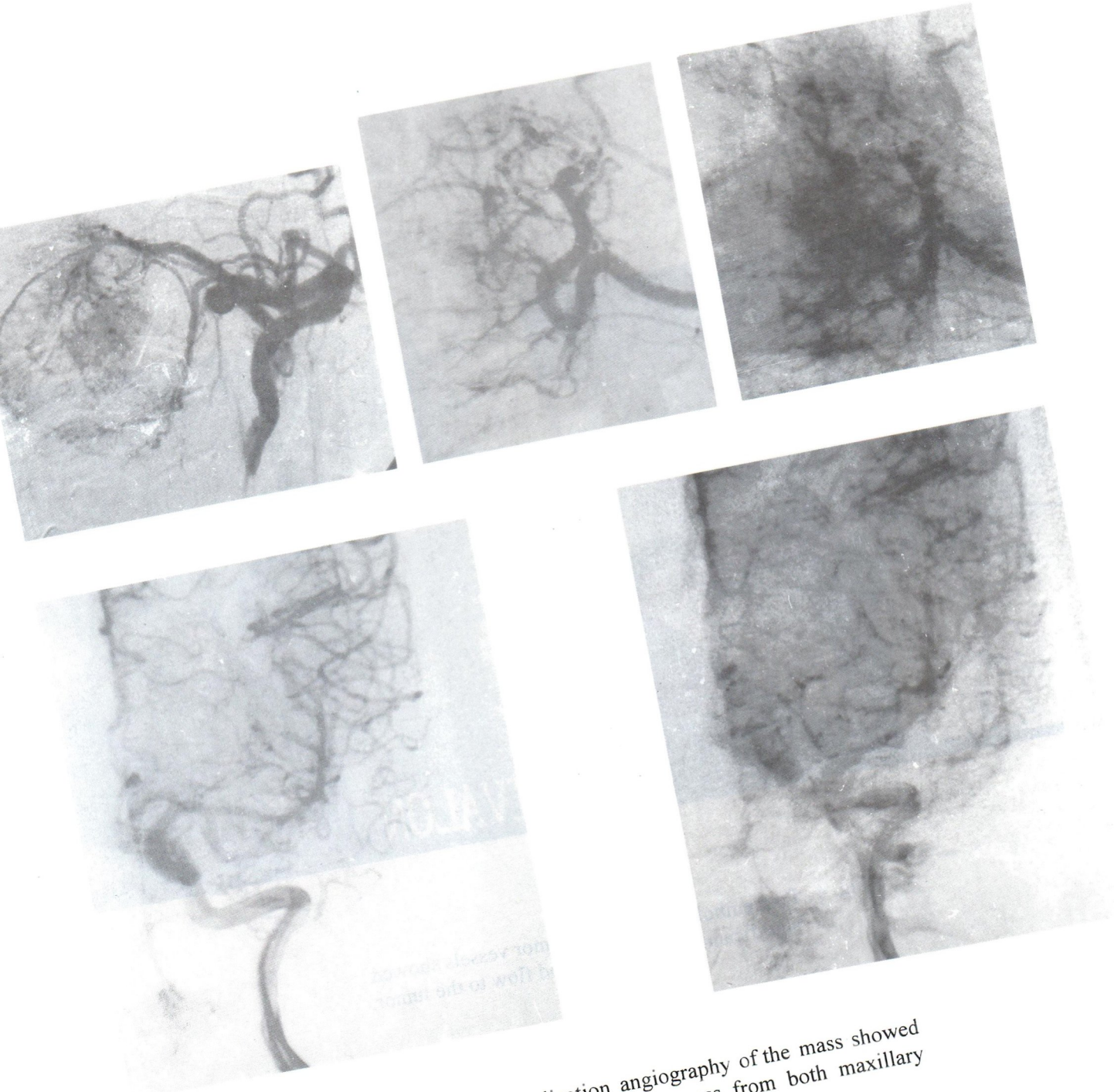

Fig.3 Preembolization angiography of the mass showed feeding arteries to the mass from both maxillary arteries and left ophthalmic artery.

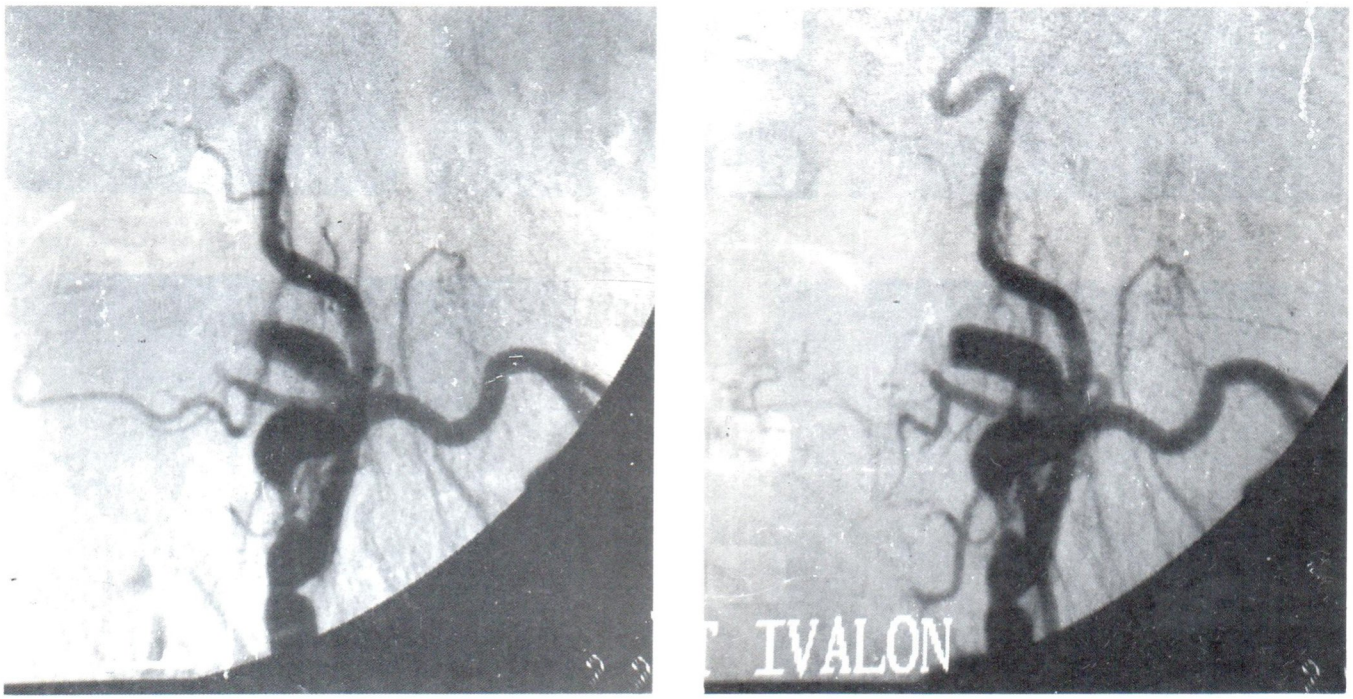

Fig. 4 Post embolization of the tumor vessels showed significantly decreased blood flow to the tumor.