

Pilomatrix Carcinoma of the Skin

—Report of a Case with Clinical Evidence of Distant Metastasis†—

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ABSTRACT

Pilomatrix carcinoma is an extremely rare tumor of hair matrix origin and its definite biological behavior of malignancy has remained inconclusive. We report a case of a huge pilomatrix carcinoma arising in the lateral neck skin from an elderly woman, who eventually developed distant metastasis to thoracic vertebra.

INTRODUCTION

Pilomatrix carcinoma is a malignant counterpart of pilomatricoma, firstly designated by Lopansri and Mihm¹⁾ in 1980, but the name of matrical carcinoma²⁾ or calcifying epitheliocarcinoma³⁾ was also synonymously used. Recent studies by light and electron microscopical observations and histochemical studies favor the hair matrix as the site of its origin, but this tumor is extremely rare compared to the benign form of pilomatricoma and only several instances have been reported in English literature under the different names¹⁻⁵⁾. It shows some histologic resemblance to pilomatricoma, but differs by the presence at the advancing edge of cytologically atypical cells without the usual shadow cell transformation and maturation as shown in pilomatricoma. As the name implies, its local recurrence and aggressive fashion with invasive growth are expected, but distant metastasis has not been reported. We report

a case of pilomatrix carcinoma with histological proof of malignant neoplasm and clinical evidences of distant metastasis.

CASE REPORT

A 64 year old woman presented with an 8-year history of a small nodular skin lesion on her right lateral neck. The lesion grew rapidly for last one year from a bean-sized nodule to a 11 cm mass with ulceration of covering skin. The tumor was widely excised under the impression of squamous cell carcinoma of the skin. Four months after excision of the tumor, paraplegia and voiding difficulty developed. Radiologic studies including myelography of thoracic spine strongly suggested a metastatic lesion in the second thoracic vertebra (Fig. 1). Physical examination and routine laboratory examinations failed to find other primary lesion than the removed mass of the neck. The vertebral lesion was irradiated with a total dose of 3,000 rads under impression of a metastatic pilomatrix carcinoma and subsequent relief of symptoms was followed. Local recurrence was not found after surgery for last one year.

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PATHOLOGIC FINDINGS

Grossly the mass measured 11×4×3 cm and involved both dermis and subcutis (Fig. 2). The covering skin surface was irregularly ulcerated, and on section the mass was tightly attached to the skin in the central portion. The tumor was multilobulaed

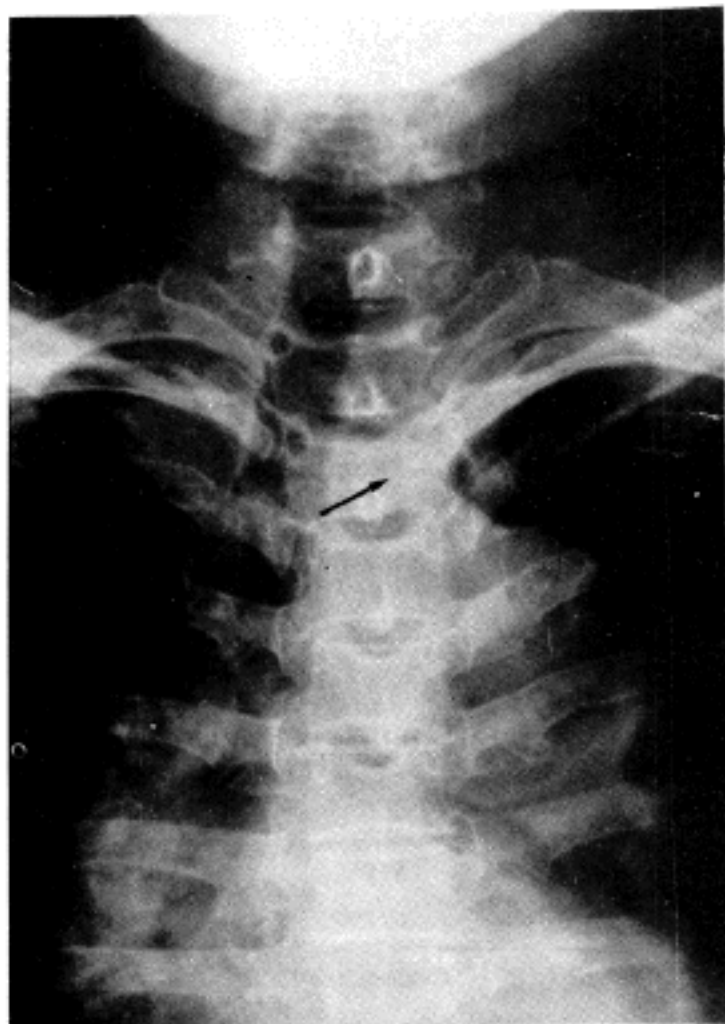


Fig. 1. Chest postero-anterior view. Decreased height of vertebral body and poorly delineated left pedicle with sclerosis of the second thoracic vertebra, highly suggestive of metastatic lesion.



Fig. 2. The cut surface of the tumor showing multilobular appearance with irregular margin and surface ulceration.

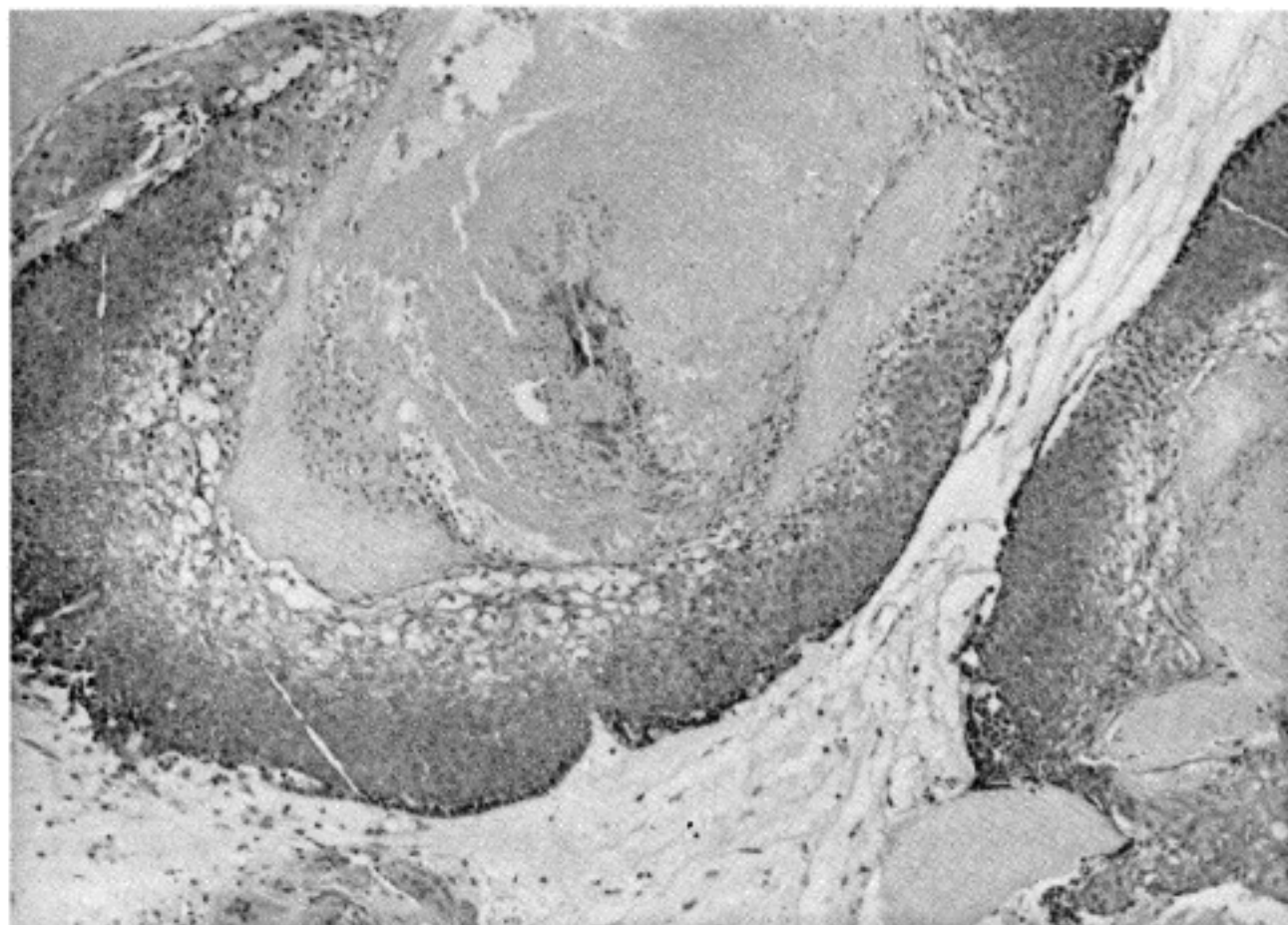


Fig. 3. Area showing typical features of pilomatricoma composed of basaloid cells, shadow cells and cellular debris. (H&E, ×100)

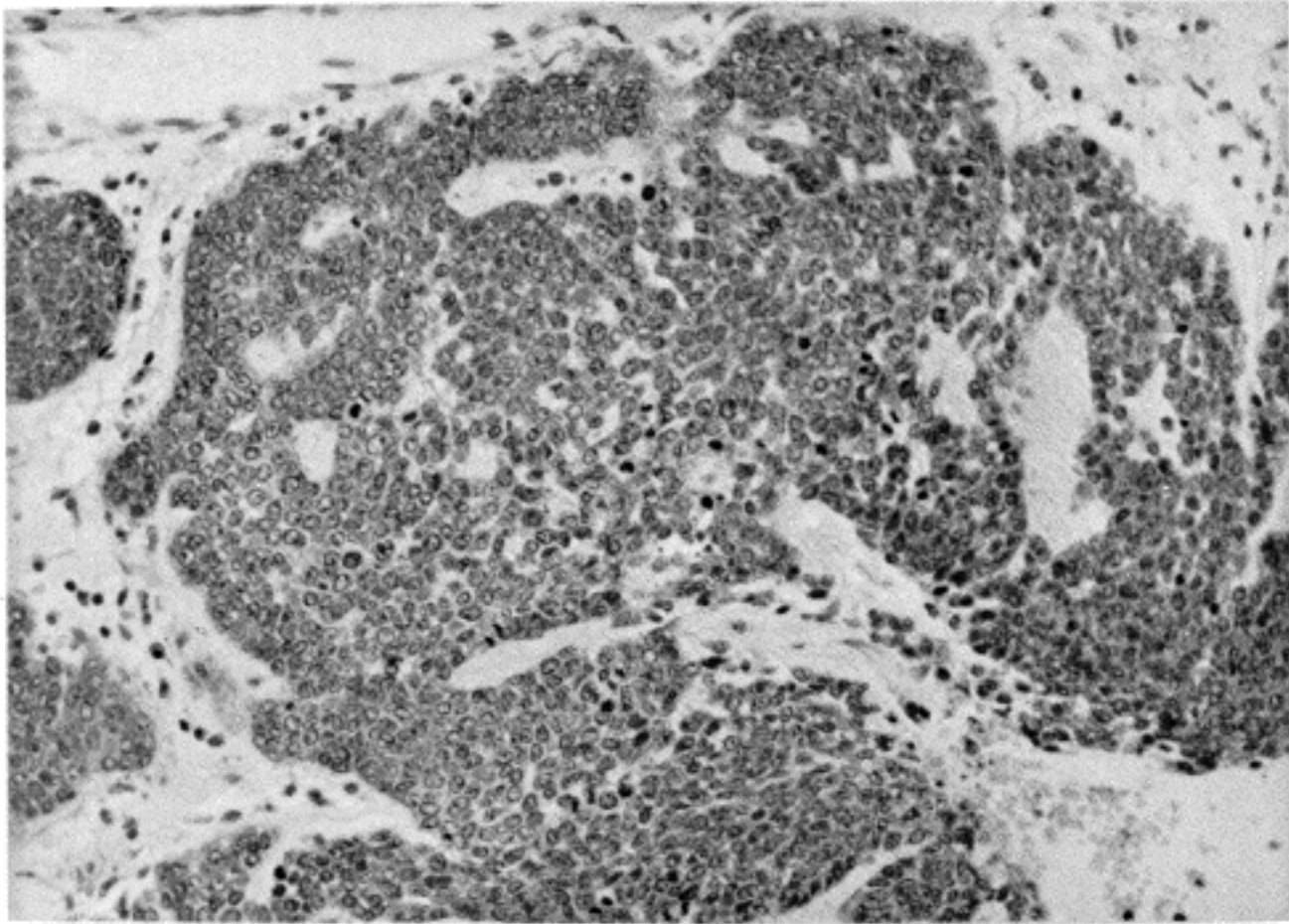


Fig. 4. Infiltrating peripheral tumor nests composed of basaloid cells with little maturation. (H&E, $\times 200$)

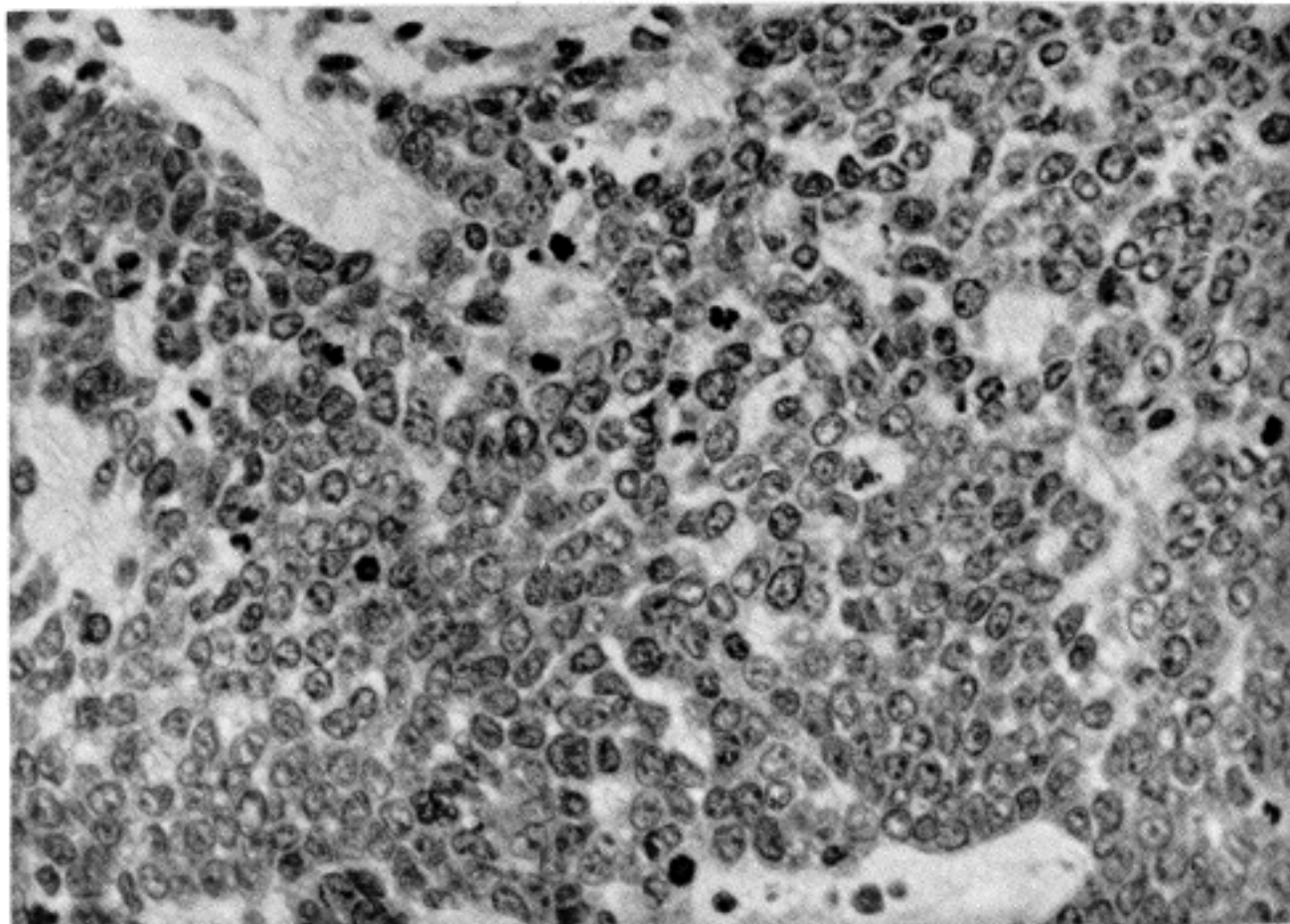


Fig. 5. Actively proliferating anaplastic basaloid cells with numerous mitoses. (H&E, $\times 400$)

and pale greyish white with widespread small yellow granular streaks, and tumor margins were sharply

demarcated from the adjacent subcutaneous and dermal tissue but without recognizable capsular

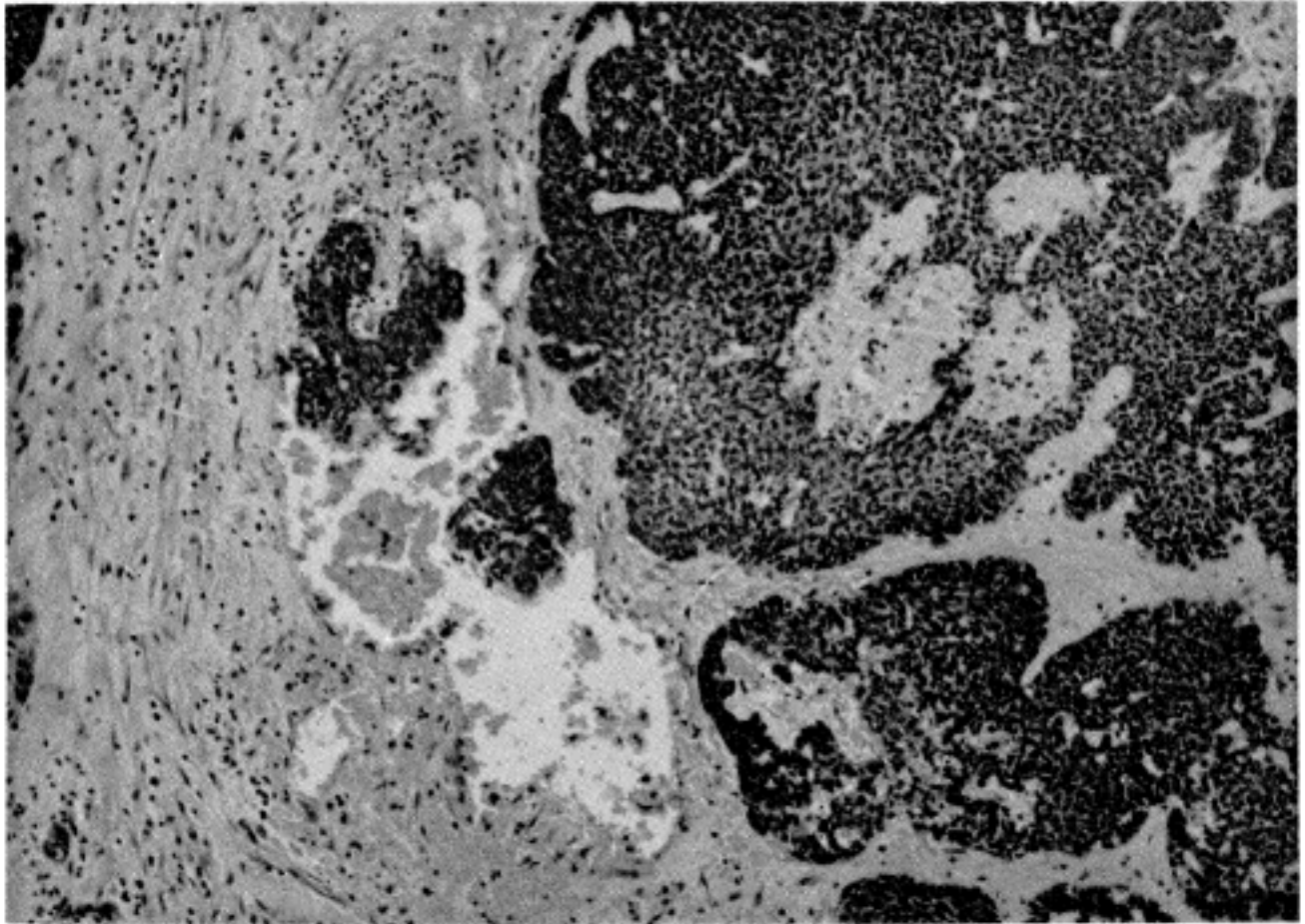


Fig. 6. Vascular invasion by basaloid tumor cells. (H&E, $\times 100$)

structure. Small discrete tumor nodules were separately found from the main mass. The consistency was firm.

Microscopically, sections showed generally aggressive tumor growth with matrical differentiation to form typical features of pilomatricoma which was composed of central keratotic material, foreign body giant cells, pale eosinophilic keratinocytes, ghost or shadow cells and amorphous debris (Fig. 3). The actively growing edges were composed of islands of numerous large, vesicular, hyperchromatic, proliferating basaloid cells with little cytoplasm (Fig. 4). No or little matrical differentiation was suggested in the solid grayish white tumor tissue, especially in the infiltrative nodules. Mitoses were frequent and numbered as high as 30/HPF (Fig. 5). Vascular invasion by the anaplastic basaloid cells was noted in the vicinity of main tumor (Fig. 6).

DISCUSSION

This pilomatric carcinoma showed more aggres-

sive features than ever reported¹⁻⁴), based on the features that ① this tumor grew rapidly for last one year from a bean sized nodule to a 11 cm mass with ulceration of surface, whereas all reported pilomatric carcinomas were slowly growing tumor without surface ulceration¹⁻⁴) and their size were much smaller ranging from 1.2 cm to 7 cm, except one case which was 13.5 cm in size; ② microscopically, infiltrating tumor cells showed more frequent mitosis with atypism than the cases reported; ③ vascular invasions by basaloid neoplastic cells were noted in the tumor; and ④ clinical evidence of distant metastasis. Although the metastatic thoracic vertebral lesion was not histologically confirmed, the vertebral lesion may represent a metastatic pilomatric carcinoma because physical and routine laboratory examination failed to detect other lesions in spite of vigorous study. Furthermore, bone metastasis is also one of the common site in many skin adnexal carcinomas⁵), and the vertebral lesion responded with radiotherapy.

While the benign lesion of pilomatricoma is one of

the common skin adnexal tumors and comprises approximately 0.1% at this institution, its carcinomatous counterpart has not been experienced so far in Korean literature. Cumulative reports will answer the more precise biological nature of this pilomatrix carcinoma.

REFERENCES

- 1) Lopansri S, Mihm MC: *Pilomatrix carcinoma of calcifying epitheliocarcinoma of Malherbe. A case report and review of literature. Cancer* 45:2368, 1980
- 2) Weedon D, Bell J, Mayze J: *Matrical carcinoma of the skin. J Cutan Pathol* 7:39, 1980
- 3) Prandetsky AP, Yuzvinkevich AK: *Malherbe's epithelioma with signs of malignization. Arch Pathol* 31:64, 1969
- 4) Sasaki CT, Yue A, Enriques R: *Giant calcifying epithelioma. Arch Otolaryngol* 102:753, 1976
- 5) Mikhail GR, Nims LP, Kelly AP Jr, et al: *Metastatic basal carcinoma. Arch Dermatol* 113:1261, 1977

— 국문초록 —

원격부 전이의 가능성을 시사하는 모기질암종 1예

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모기질암종은 극히 드문종양으로서 모기질종의 악성형이라고 이해되고 있으며 국소성 침윤성 성장과 제거후의 흔히 재발을 특징으로 하나, 전이했다는 보고는 아직 없다. 저자들은 전이의 가능성이 있는 모기질 암종 1예를 경험하고 보고한다.

종양은 64세 여자의 우측 경부에서 1년사이에 콩알크기로부터 11 cm 크기로 빨리 커졌고 궤양을 동반하였다. 조직학적으로 종양부는 전형적인 모기질종의 특성을 보이나 주변부는 분화가 되지 않은 기저양세포로 이루어져 있고 핵분열은 대단히 많아서 한 고배율시야에서 30개 까지 보였으며 종양세포의 혈관침입도 관찰되었다.

이 종양을 제거하고 4개월뒤에 제 2요추에 전이성 병변이 발견되었는데 이 병변은 방사선 치료에 잘 반응하여 증상의 호전을 초래하였다. 요추의 전이성병변에서 생검이 시행되지는 않았으나 이학적검사와 기타 검사로서 다른 원발성 종양이 있다는 증거가 없어서 요추의 병변은 경부의 모기질 암종이 전이한 것이라고 추정하였다.