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Rhabdomyoma of the Orbit

—A report of a case—

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Extracardiac rhabdomyomas are rare benign neoplasms of skeletal muscle origin. Particularly, orbital rhabdomyoma is a very rare entity in spite of relatively common occurrence of its malignant counterpart, rhabdomyosarcomas. In world literature, 6 cases of orbital rhabdomyomas had been described until 1975. Among them, earlier 5 cases are not readily acceptable now owing to the recent understanding in the diagnosis of extracardiac rhabdomyoma.

Rhabdomyomas can be subdivided again into fetal and adult type. The adult type is a benign tumor comprising of mature but disorganized striated muscle fibers. Although called "adult type" this well differentiated type of rhabdomyoma may develop in childhood. There has never been an instance of rhabdomyosarcoma arising in, or associated with, a rhadbomyoma. Therefore, it is essential that these neoplasms be recognized histologically and that they be treated conservatively by local excision.

CASE REPORT

The patient was a 7 months old Korean girl, who was admitted to Department of Ophthalmology of the Seoul National University Hospital, with 6 months history of the left eyelid swelling and exophthalmos. Visual acuity and ocular movement were unaffected. Orbit CT revealed 1.5×1.5 cm sized well circumscribed intraconal mass in the left retrobulbar space with pressure erosion of adjacent bone. At surgery, the mass was pinkish gray and firm, and was adherent to adjacent tissue. The whole mass could not be removed, and a local excision was done.

PATHOLOGIC FINDINGS

Grossly the tissue excised for biopsy consisted of 5 fragments of grayish white soft tissue, measuring 5 cc in total volume. They were fixed in 10% formalin solution. Histological and electron microscopic examinations were done. Histologically the tumor was composed of compactly arranged large round or oval to polygonal cells and some fibers. Their cytoplasm was eosinophilic and finely granular with occasional vacuoles which stained positive on PAS staining. Thin fibrous septa were seen traversing this rather cellular tumor containing scanty stroma. Some cells showed definite cross striations which were more obvious on PTAH and Masson trichrome staining. The nucleus was large, hyperchromatic, and vesicular. It contained one or two prominent nucleoli. Mitotic figure

was not seen anythere.

Electron microscopically, the cytoplasm of the tumor cells showed organelles characteristic of skeletal muscle cell although they lacked the normal arrangement. The mitochondria were numerous and of various shape and size. Some cells showed orderly arrangement of myofibrils with well preserved cross striations. Most of cells revealed irregularly arranged and aggregated thin myofilaments of varying length. Isolated rod-like structures of hypertrophied Z-band material were noted. A-bands and I-bands were not identified.

DISCUSSION

The earliest cases of orbital rhabdomyoma were described by Jennings[10], Alt[2] and Wible[20]. Calhoun and Reese[5] regarded these tumors as rhabdomyomas of mature form in their review of myogenic tumors of the orbit. Since then, Forrest[8] and Nath and associates[16] reported additional 2 cases, describing them as 4 th and 5 th case of striated muscle tumor of the orbit. In 1975, Knowles and Jacobiec[13] described another case which had been discovered during the review of orbital rhabdomyosarcomas in their file, as the first to be documented. They concluded that the previous five cases lacked adequate clinical, descriptive and photographic documentation to be unequivocally accepted as rhabdomyomas. The tumor, which they reported, was composed of compactly arranged large, round to polygonal cells with abundant granular acidophilic cytoplasm and single eccentrically placed large vesicular nucleus containing prominent central basophilic nucleolus. They observed many cells containing glycogen vacuoles with spider cell formation, cells showing well-organized cross striations and some cells having multiple acidophilic rod-like crystals in their cytoplasm. These findings and the presented photographs are identical with the first description of extracardiac rhabdomyoma by Moran and Enterline[15] and those of further cases of nonophthalmic origin. In comparison, among the earlier cases of recorded orbital rhabdomyoma, at least those of Forrest and Nath and associates are most likely to be inflammatory pseudotumors in regard of inflammatory infiltrate and fibrous proliferation. Those of Jennings, Alt and Wible afford little information about the detail histopathology.

A spate of case reports with ultrastructural study has helped to characterize this tumor, which is now divided into adult and fetal type. The adult type rhabdomyoma, which we discuss here, almost exclusively occur in head and neck region, particularly larynx and pharynx of adult male. To the best of our knowledge the present case and that of Knowles and Jacobiec are the only adult type rhabdomyoma that occurred in the orbit of children. The present case however is slightly different from Knowles' case. Spider cells were not observed in our case, and PAS stain revealed only small amount of scattered glycogen. Glycogen glanules were also not abundant on electron microscopy either.

Intracytoplasmic crystalline structures, which were discribed as myosin crystals by Knowles and Jacobiec, now thought to be hypertrophied Z-band, were not definite on light microscopy. However, electron miscroscopy revealed haphazardly arranged abundant myofilaments and isolated aggregations in most of cells which showed granular cytoplasm without demonstrable cross striations on H-E stain. Though our EM study was not sufficient due to small amount of paraffin-embedded

preparation, these electron and light microscopic findings are consistent with adult type rhabdomyoma. And we document the present case of rhabdomyoma occurred in orbit.

SUMMARY

A case of adult type orbital rhabdomyoma is reported. This 7 months old girl had a left retroorbital tumor of 1.5cm diameter, which showed characteristic histological findings of adult type rhabdomyoma. Light microscopy revealed compactly arranged round to polygonal cells having abundant eosinophilic granular cytoplasm and large vesicular nuclei. Glycogen vacuoles and cross striations were occasionally observed. Electron microscopy revealed irregularly arranged and aggregated thin myofilaments in the cytoplasm of most cells. Isolated structures of hypertrophied Z-band were also demonstrable. Occasionally, regular myofilaments showing cross striation were observed.

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=국문초록=

안와의 횡문근종

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심장외에서 발생하는 횡문근종은 대단히 드문 양성 종양으로서 특히 어린이 안와에서 발생하는 경우 그 임상상이나 조직상이 악성 종양과 감별을 필요로 한다.

본 예는 7개월 여아에서 발생한 안와종양으로서 생후 1개월부터 좌측안검의 부종과 안구돌출증이 있어 7개월에 입원 수술하여 안구뒤에 있는 종괴를 부분 절제한 예로서 병리조직학적으로 횡문근세포로 구성된 양성종양으로서 치밀하게 배열된 크고 둥글고 호산성 세포질을 가지는 횡문근 세포로 구성되어 있었고 핵은 크고 농염되었고 흔히 편위되어 있었다. 이러한 조직학적 유형은 이른바(성인형) 횡문근종의 소견에 합당하였으며 전자현미경 검사상 풍부한 myofilament와 과형성의 Z-band를 보였다.

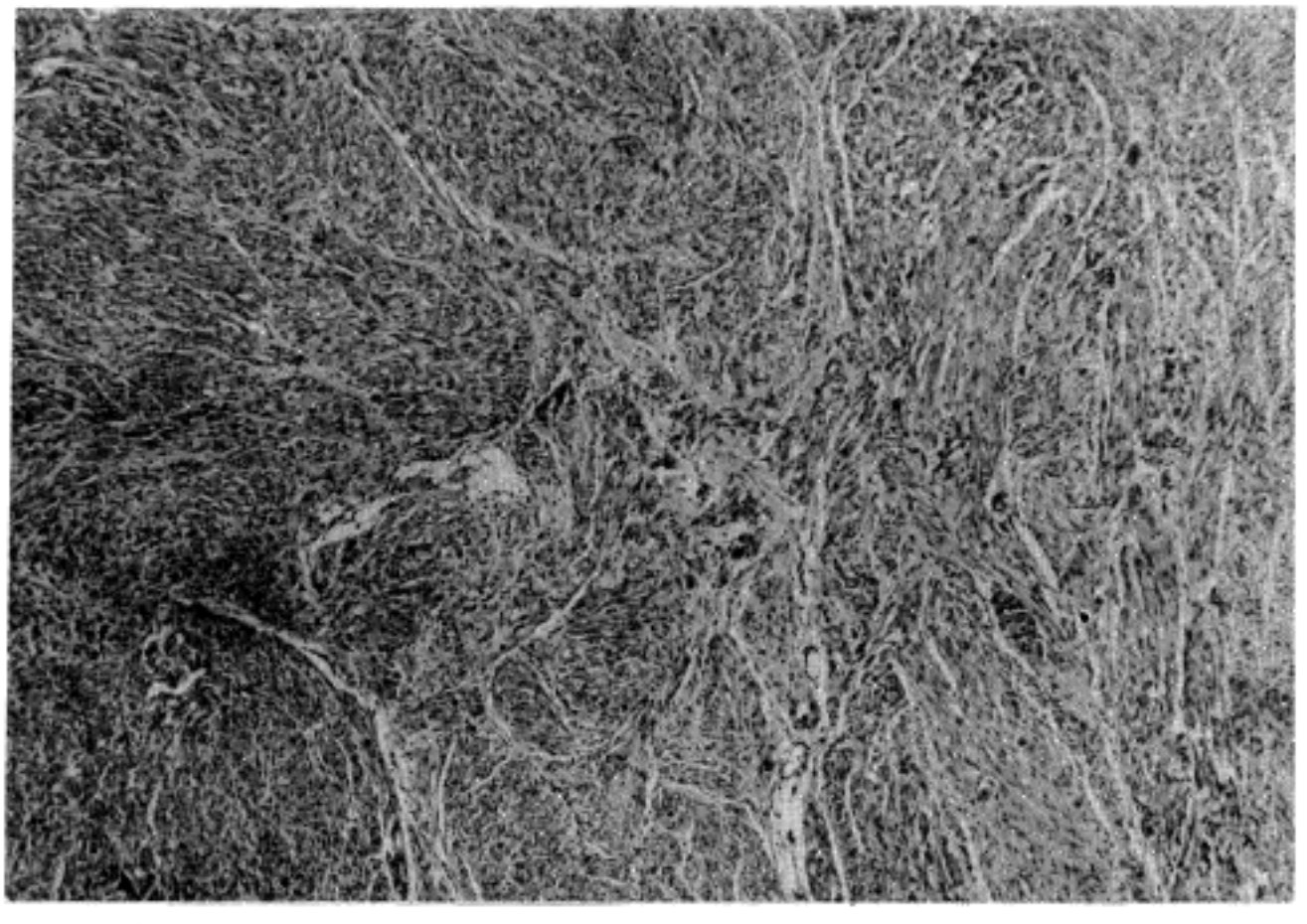

Fig. 1. The tumor is composed of compactly arranged oval to polygonal cells with traversing loose connective tissue septa. (H&E, ×40)

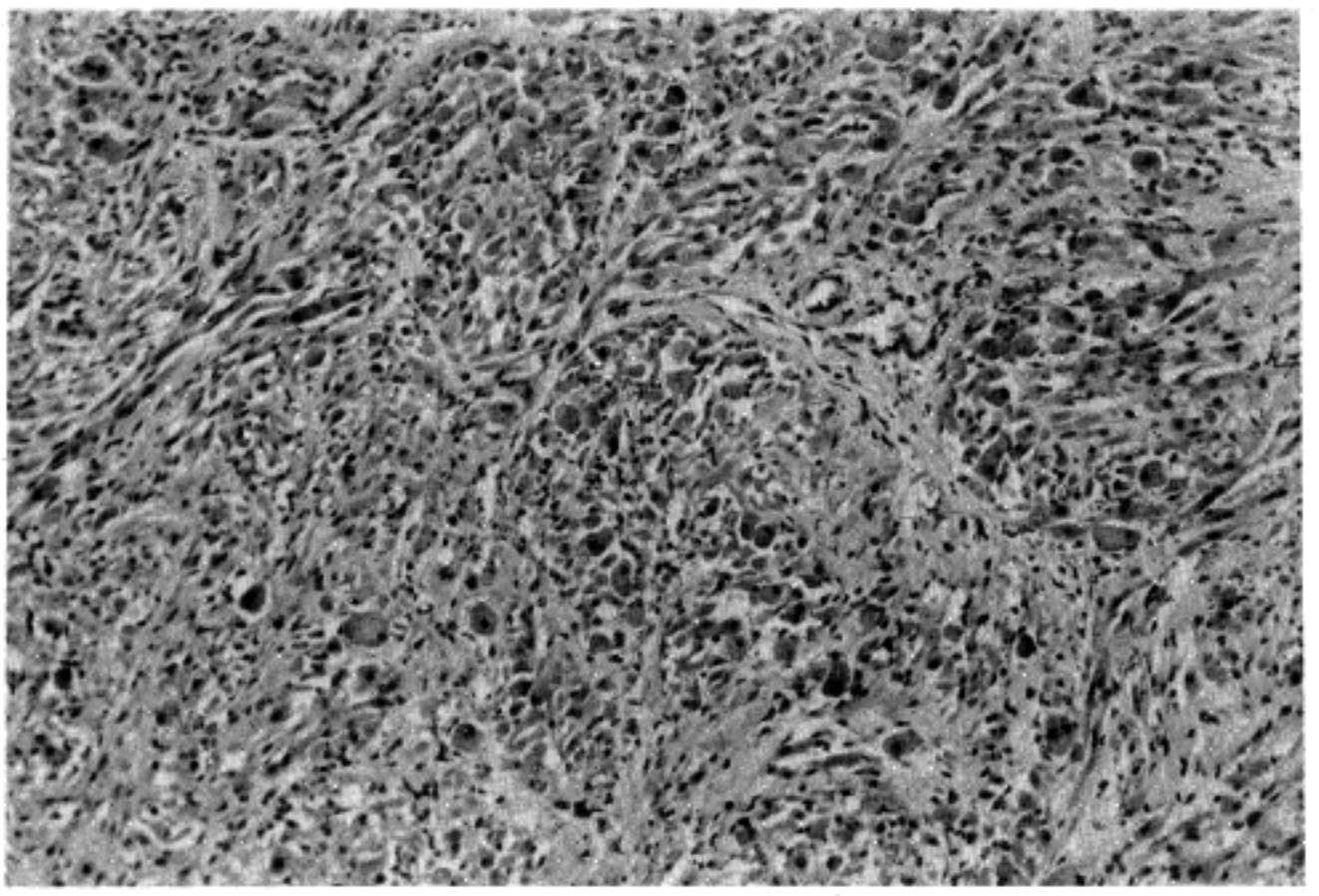

Fig. 2. The cells have abundant eosinophilic cytoplasm and eccentrically placed large hyperchromatic nuclei. (H&E, ×100)

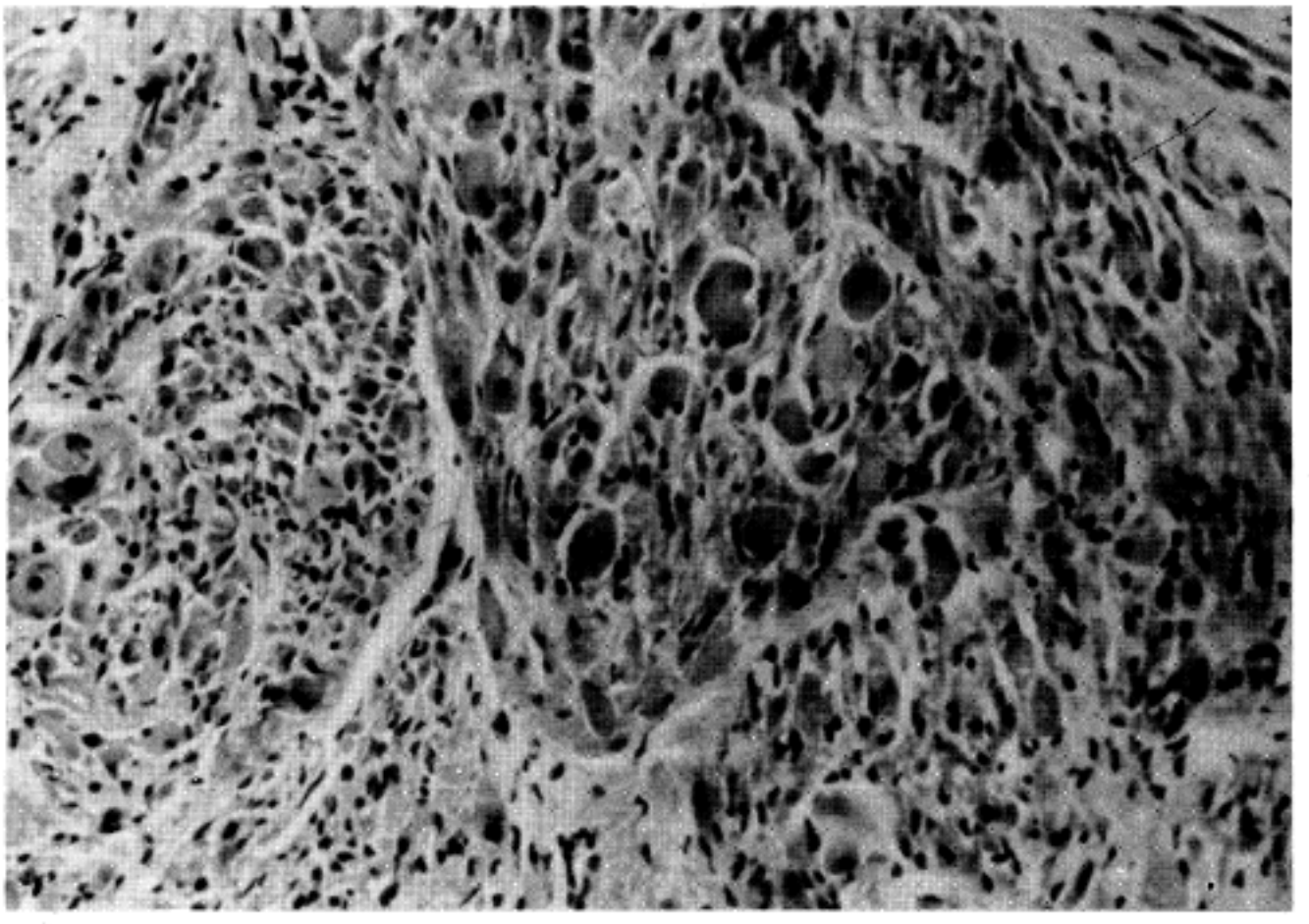

Fig. 3. The cytoplasm is granular and contains occasional vacuoles. (H&E, ×200)

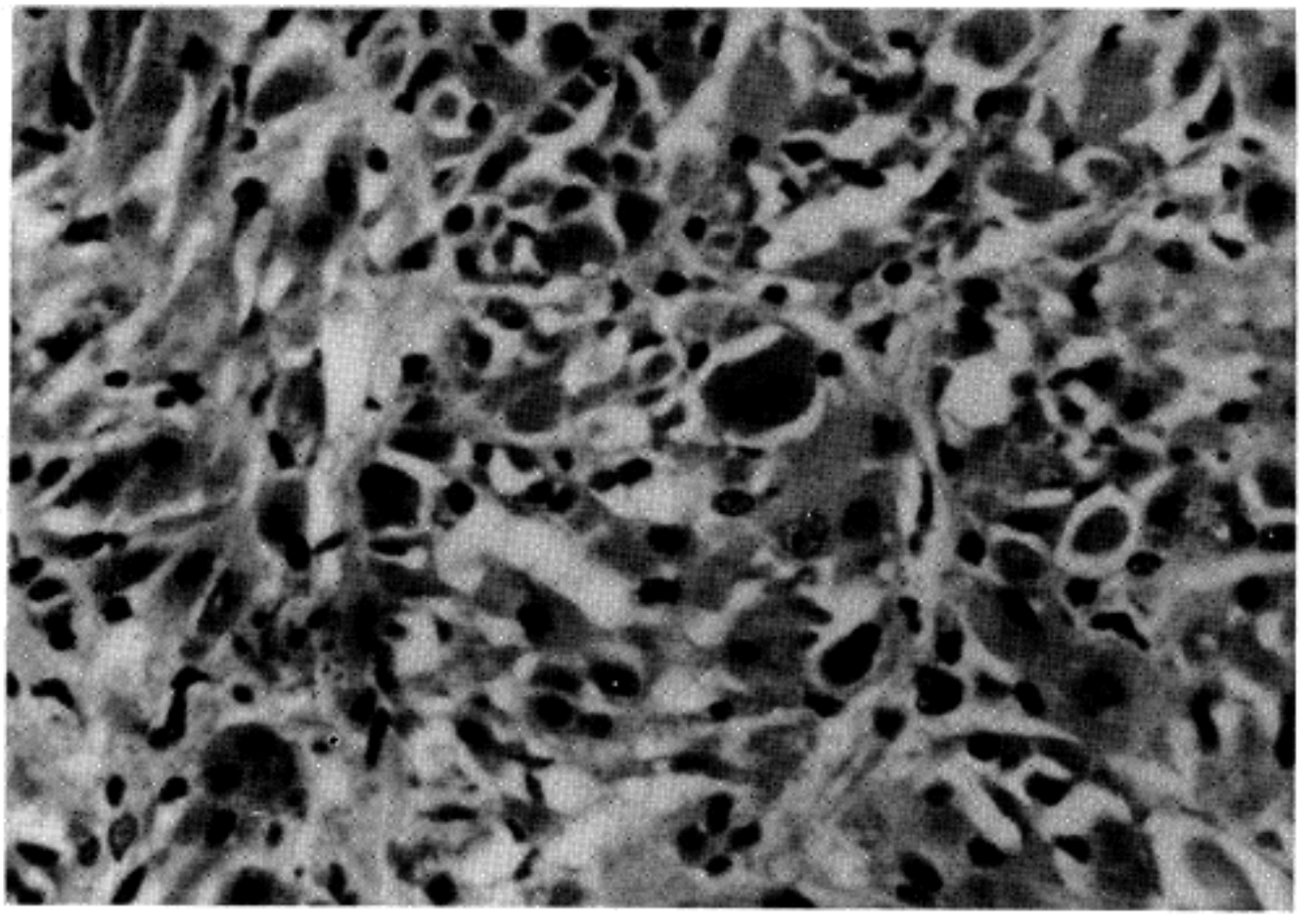

Fig. 4. The nuclei are frequently vesicular with prominent nucleoli. (H&E, ×400)

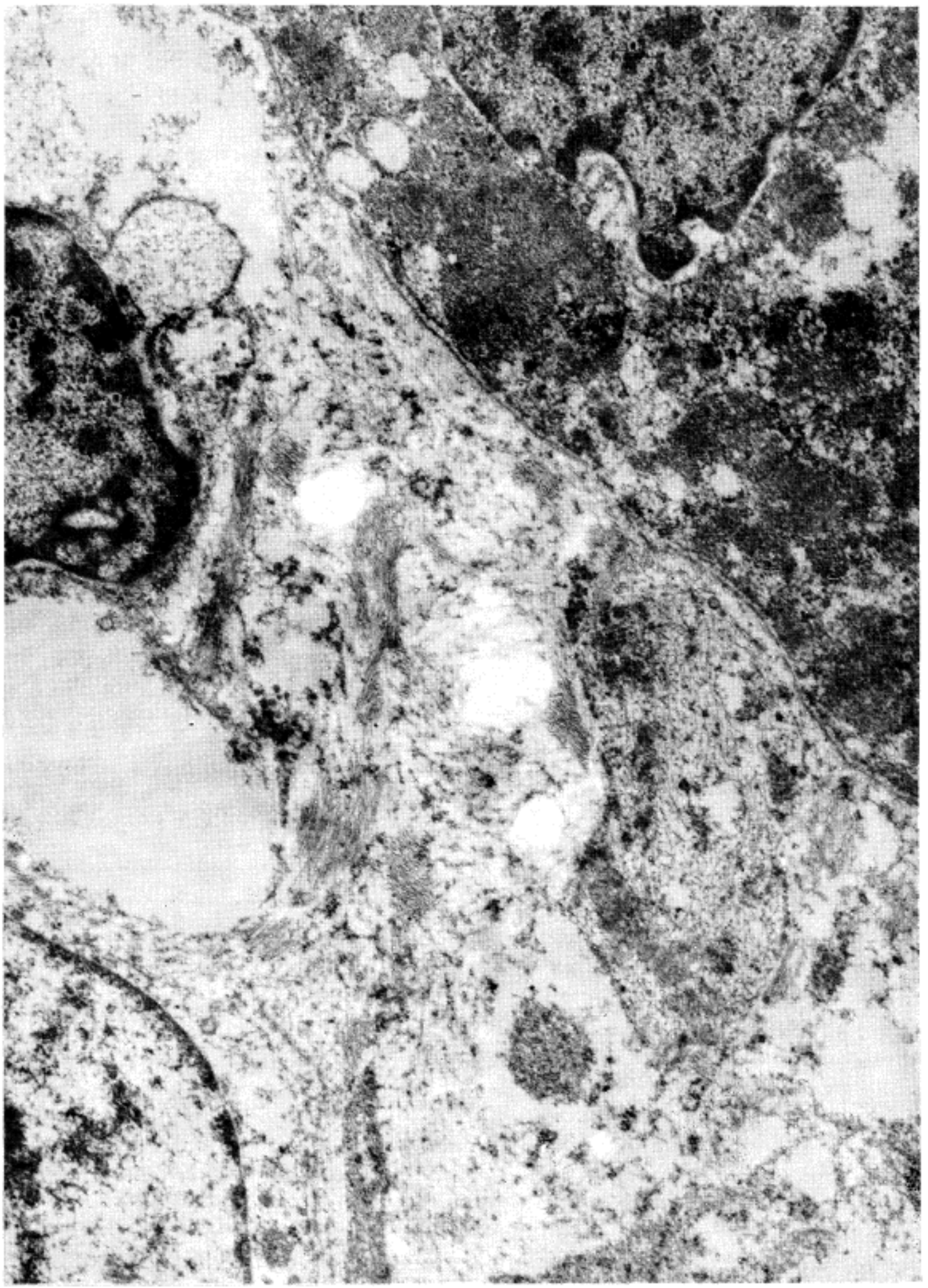

Fig. 5. The cytoplasm contains irregularly arranged numerous myofilaments. (EM, ×22400)