

CASE 7

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This man was 53 years of age at the time of death. His neurological illness began 18 months earlier when he developed pain and paresthesias in the right face following the excision of a 4 cm preauricular encapsulated mass. Eventually, he developed right lateral rectus palsy, right jaw weakness, progressive left hemiparesis, hypalgesia, difficulty in swallowing and speaking. Severe, persistent headaches developed before he expired with bronchopneumonia.

During his adolescence this individual had severe acne which was treated by irradiation. Since he worked then as a helper in a physician's office, allegedly he also gave himself facial X-ray treatments. Chronic radiodermatitis developed. During the last 10 years, many basal cell carcinomas and senile keratoses were removed from his face. Five years before the patient's demise a spindling malignant tumor was removed anterior to the right ear. One year later, this recurred. Subsequently similar lesions were found in the right upper lip, right temporal, right preauricular areas, and then also from the left lower cheek area. The basic characteristics of all these spindling tumors are similar to our diagnostic slide preparation.

Examination of the right fifth nerve at autopsy showed it to be greatly enlarged along its course into the ventral pons, where it stopped abruptly. The left fifth nerve showed no abnormality.

Submitted are: One Kodachrome transparency, 1 slide stained with H & E and 1 unstained slide.

Our interpretation is that this is a neurofibrosarcoma which developed possibly as a result of an excessive quantity of irradiation.

Questions raised by this case are:

- 1) Is there a relationship between the irradiation and the development of this malignant appearing peripheral nerve tumor?
- 2) What is the nature of the tissue alteration which induces such neoplasia?
- 3) What factors might account for the long delay before the clinical appearance of these skin lesions?
- 4) There was no evidence of von Recklinghausen's disease in this individual. Would this alter one's opinion regarding the diagnosis?