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EPIDEMIOLOGY OF ESTHESIONEUROBLASTOMA.

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Esthesioneuroblastorna is an uncommon and aggressive neoplasm arising from the olfactory epithelium of the nasal vault. A previous comprehensive literature review from 1924 to 1994 *(Broich G, Pagliari A, Ottaviani F: Esthesioneuroblastoma: a general review of the cases published since the discovery of the tumour in 1924. Anticancer Res 17: 2683-2706, 1997)* revealed a total of 945 true new cases reported in 70 years. Underdiagnosis and unreported cases can be supposed, since the tumor offers controversial diagnostic problems and definitive diagnosis requires in dept study with methods non always readily available to all centers At light microscopic examination tumor cell nests are separated by fibrous connective tissue with Homer-Wrigh rosettes and/or Flexner rosettes on an acidophilic fibrillary background. Ultrastructurally there are neurosecretory granules and neurofibrils in the cytoplasm of tumor cells. Immunocytochemical reactions show positivity to neuron specific enolase (NSE), chromogranin (CHR)7 S-100, vimentin and keratin. The tumor can be staged according to one of the classifications proposed by Kadish, Morita, Biller, or Dulgerov on the basis of physical examination, magnetic resonance (MR) images and computed tomographic studies at initial examination. Treatment schemes include radiation alone, combined preoperative or postoperative radiation and surgery, surgery alone, chemotherapy, chemotherapy and stereotaxic proton beam radiotherapy and high dose chemotherapy followed by autologous bone marrow transplantation for various stages of disease. The authors

have reviewed the literature from their original paper up to June 1998. The total of all reported cases has risen by 298 new cases from 1457 in 1994 to 1755. The true new cases rise by 75 from 975 in 1994 to 1020 now. We see that the gross yearly report rate in the literature has risen by 197.6 % from 28.6 cases/year in the period 24-94 to 85.1 cases/year in the period January 95- June 98, denoting a rise in interest. The reports of truly new cases perhaps have have risen orly from 18.5 cases/year to 21,4 cases/year (15.7%). It is obvious that mairlly new review and report cases have been discussed. Sex ratio has remained unchanged and still most reported cases are in Kadish class C (51.1% 24-98, of which 49.3% in the period 24-94 and 56.3% in the period 95-98). More analytic results will be discussed, as well as evolution of treatment schemes. We suggest the establishment of a world registry of ENB in order to avoid double reporting and favor statistical treatment of the data of this rare tumor, connected with reference centers for histological diagnosis and elaboration of the prognostic data of treatment protocols.