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ESTHESIONEUROBLASTOMA.

A Review of the data published from 1924 to 1994.

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Esthesioneuroblastoma (ENB) is a rare tumour arising from the neuroepithelium in the olfactory rim of the nasal cavity accounting for about 3% of all intranasal tumours. It was first described by Berger and Luc in 1924 as esthesioneuroepithelioma. The tumour has always been considered a rare and exotic entity, reports in literature are inconsistent and many cases are not properly accounted for. Reviews do never report more than a hundred cases, stressing the rarity of the tumour. However a thorough literature review revealed that since the first description seventy years ago, 945 true new cases have been reported.

In our search we found a total of 1,457 cases chronicled in the literature of which 487 were previously documented. Thus we estimate 945 true new cases of ENB. Author cases account for 198 and collaborative institutions for 747 cases. Sex distribution was 53.36% males and 46.64% females. Kadish classification was applied to 553 cases revealing 103 (18,29%) class A cases, 182 (32,33%) class B and 278 (49,38%) class C cases. This distribution is generally stable through the decades. Treatment could be classified in 898 cases. It consisted in Surgery alone in 25,17% (226 cases), Radiotherapy alone in 18,37% (165 cases), combined surgery and radiotherapy in 43,21% (388 cases) and chemotherapy in 13,2% (119 cases), followed in 11 cases (1,22%) by bone marrow transplant. In the reported cases an overall follow up could be valued in 477 cases, while only in 234 cases a five year follow up was done. The outcome was 68,38% alive and disease free, 12,82% alive with disease and 18,80% dead. From these 20,51% had only surgery, 11,11% radiotherapy and