Horner’s Syndrome In Infants And Children – Should We Routinely Screen For

Neuroblastoma ?

Background

Horner’s syndrome (HS) – ocular miosis, ptosis and anhidrosis – may be a subtle sign of occult pathology in otherwise asymptomatic children, neuroblastoma (NBL) being the commonest associated malignant tumour. Despite such knowledge robust evidence to guide best clinical practice is sparse and the incidence of occult malignancy in children with Horner’s syndrome remains largely unclear. We performed a systematic review aimed to ( 1 ) identify the incidence of neuroblastoma in children with HS of unknown aetiology, and ( 2 ) establish if screening for neuroblastoma should be routinely performed in this patient population.

Methods

Systematic review of the literature (PubMed and Ovid/ Medline database, 1961-2018).

Results

Initial search identified 334 manuscripts; 269 records were screened after duplicate removal. Reasons for exclusion: paper does not address research question, not English language, case reports, review or comment. Six papers were included in the final analysis. All reports were single-centre retrospective studies without control groups.

All studies included patients with unilateral miosis +/- associated signs with or without positive cocaine test, without previously known diagnosis. Studies included a total of 137 patients ( age range 0-20 years ; mean age 3.32 years).

The overall incidence of a lesion detected was 12.4% (17/137). Neuroblastoma was diagnosed in 7.3% (10/137). Seven other patients were diagnosed with - rhabdomyosarcoma ( n=1) , Ewing’s Sarcoma (n=1), ganglioneuroma ( n=1 ) , astrocytoma (n=1), juvenile xanthogranuloma ( n=1), intraorbital hemolymphangioma (n=1) and brain stem vascular malformation( n=1 ).

Conclusion

It is well recognized that HS may be the first sign of an occult malignancy, neuroblastoma being the most common one. We report the first systematic review study that comprehensively investigates the incidence of occult malignancy in this unique patient cohort. The study crucially shows that suspicion of HS of unknown aetiology in children should involve further investigation(s) to exclude an underlying space occupying lesion. We strongly recommend CT MRI imaging of the brain neck and thorax , plus urinary catecholamines for prompt diagnosis and treatment..