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# Polycythemia with Cerebellar Hemangioblastoma

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*Keywords:* *polycythemia, cerebellar hemangioblastoma.*

*GJMR-K Classification:* *NLMC Code: WL 320*


*Strictly as per the compliance and regulations of:*

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## I. Case Details

56 year old male presented with vomiting, upper abdominal pain of ~30 days duration, associated with mild weight loss; he was a known hypertensive on irregular treatment but not a diabetic; he was a non smoker, non ethanol consumer, he used snuff, his wife had undergone permanent sterilization. General examination showed medium built middle aged male, with moderate dehydration, but no pallor, icterus, lymphadenopathy or pedal edema; his blood pressure measured 150/100 mm Hg; cardiovascular system, respiratory systems showed no abnormality; he was conscious oriented, with no neurological deficit; epigastrium showed tenderness, without guarding or rigidity; there was no hepatosplenomegaly.

His initial hemoglobin was 18gm%, with hydration the hemoglobin reduced to 16.5 to 17 gm%; liver function tests showed mild elevation of transaminase to 42 mg/dl, renal parameters were normal; peripheral smear showed reactive lymphocytosis, normochromic, normocytic red blood cells, with normal white blood cells, adequate platelets. Ultra sonogram of the abdomen showed simple renal cortical cysts, fatty change liver. Oesphago gastro duodenoscopy showed pan gastritis; bone marrow biopsy done to evaluate for reactive lymphocytosis, with suspicious borderline polycythemia, showed hyper cellular marrow with erythroid, megakaryocytic hyperplasia.

He received treatment with proton pump inhibitors, parenteral hydration; patient`s vomiting stopped, his abdominal pain improved and he requested discharge due to personal reasons.

6-7 months later patient returned with weakness, debility, for the past >30 days duration with obvious weight loss; examination suggested subtle Cerebellar signs, with suspicious, mild nystagmus on looking to the left side, suspicious impairment of tandem walking, there was no other neurological deficit; investigations revealed hemoglobin of 19.5 to 20 gm%, hematocrit of 56.1%, other cell lines were normal; the weight loss was very significant ~ 50%; since true secondary polycythemia was suggested by pure erythrocytosis without involving the other cell lines, ultra sonogram abdomen was normal, with significant weight loss, computerized axial tomography scan of brain was planned, to assess for Cerebellar Hemangioblastoma, probably having resulted in secondary polycythemia.

Computerized axial Tomography scan of the brain showed a 3.5cm × 3.8 cm mass lesion, with cystic areas, suggesting a possibility of Hemangioblastoma, with complete effacement of 4$^{th}$ ventricle, seen to cause obstructive hydrocephalus of the 3$^{rd}$ and both lateral ventricles; retinal examination showed papilloedema, but no hemangiomas; Magnetic resonance imaging of the brain showed solid, cystic mass in vermis, left Cerebellar hemisphere, to cause obstructive hydrocephalus Figure 1.

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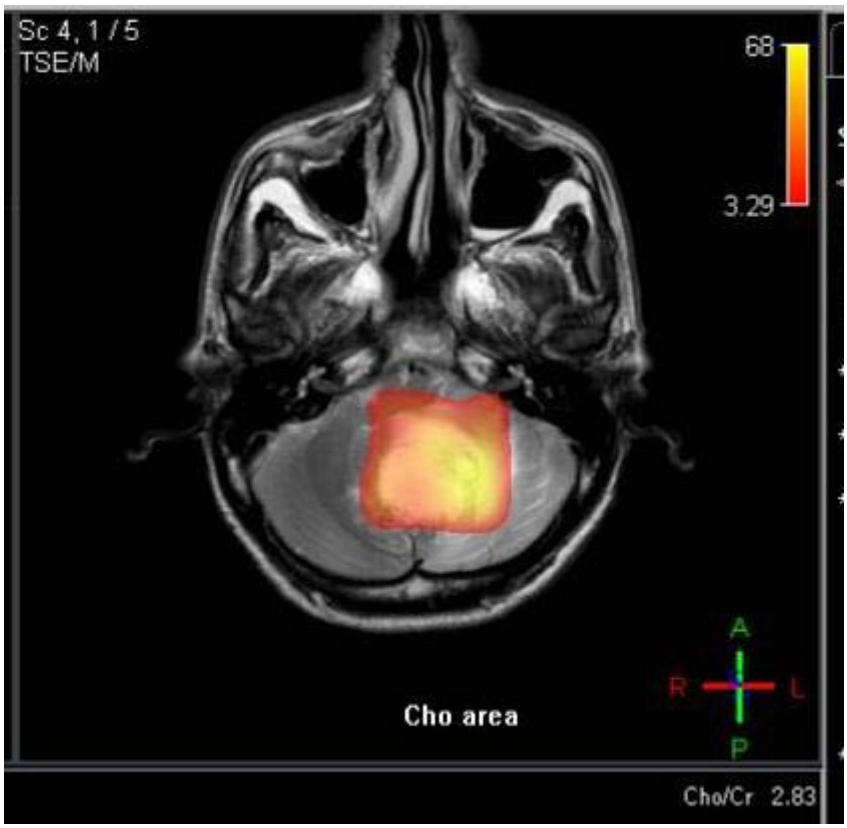
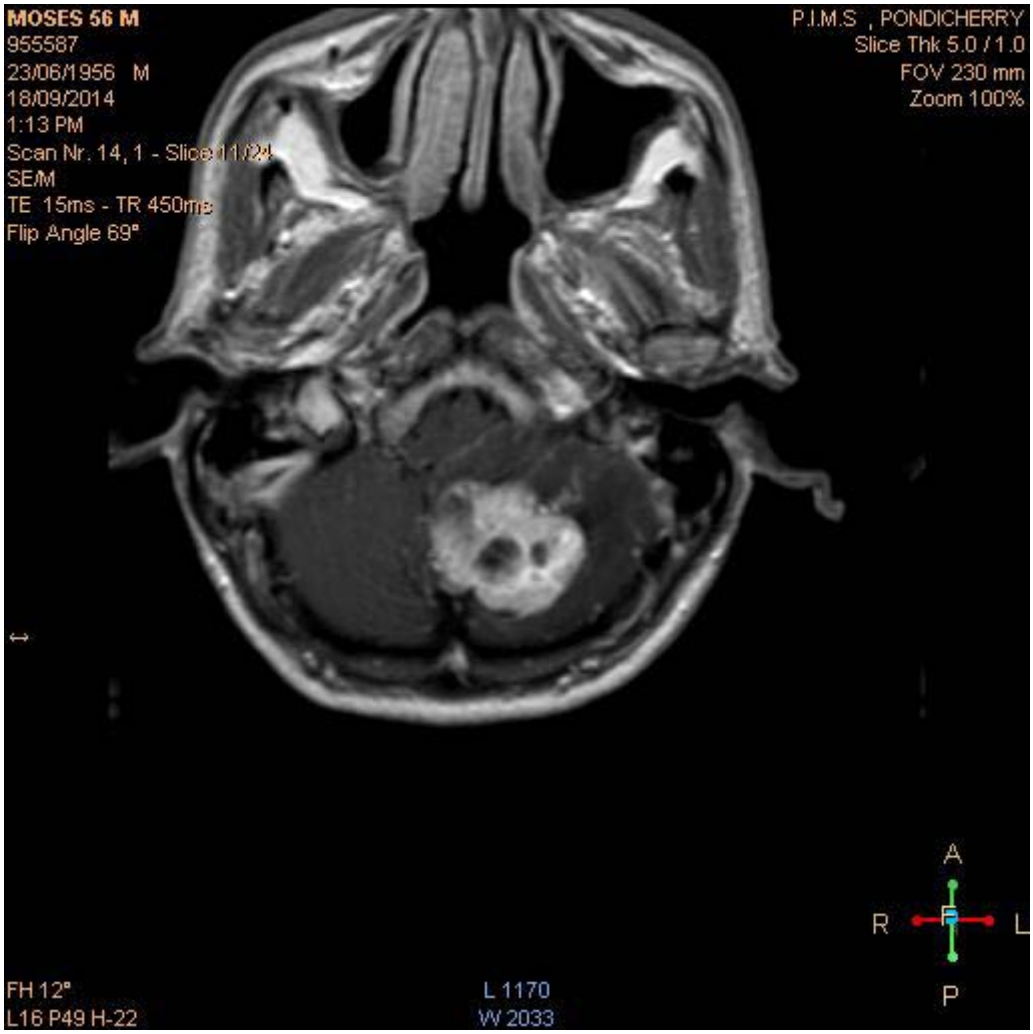



*Figure 1:* *Tumor in cerebellum*

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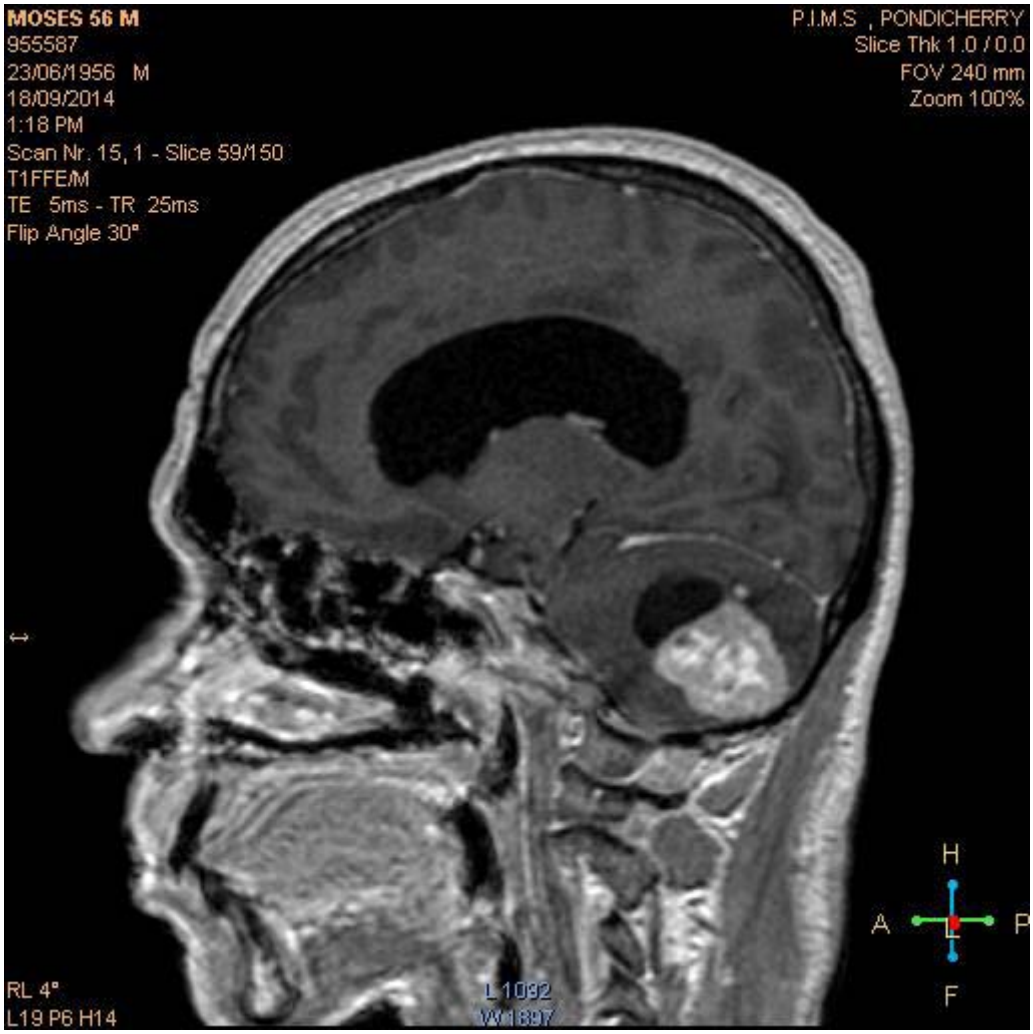



*Figure 1 :* Obstructive hydrocephalus caused by the tumor.

Emergency ventriculo peritoneal shunt was done followed by tumor excision by sub occipital craniectomy; post operative period was uneventful;

Histopathological examination of the excised tumor showed two components of the tumor-vascular and stromal; vascular component is in the form of network of capillaries and large thin walled vascular spaces; stromal component showed vacuolated large stromal cells with round to oval bland nuclei; occasional enlarged nuclei were seen; sparse mitoses were seen; foci of hyalinized vascular stroma were noted; tumor parenchyma interface was discrete in many foci, while creeping tumor was seen in occasional foci; capillary Hemangioblastoma-WHO grade 1 was reported Figure 2.

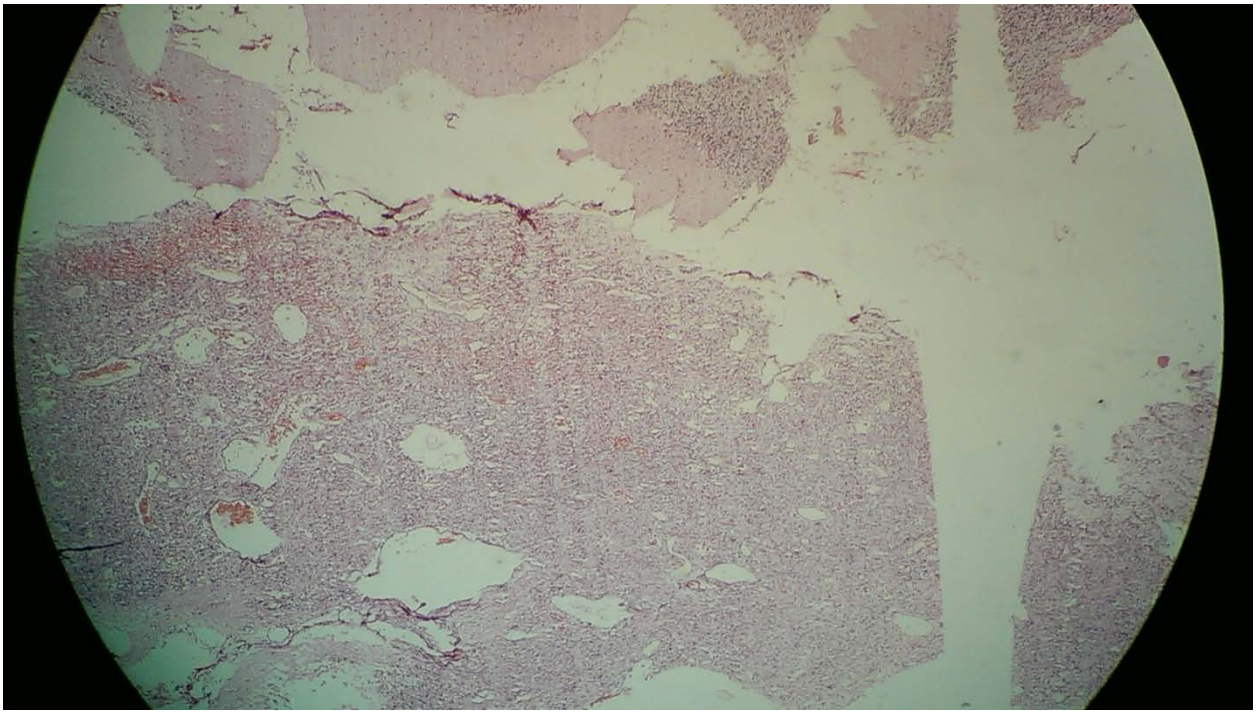


*Figure 2 :* HPE-Cerebellar Hemangioblastoma

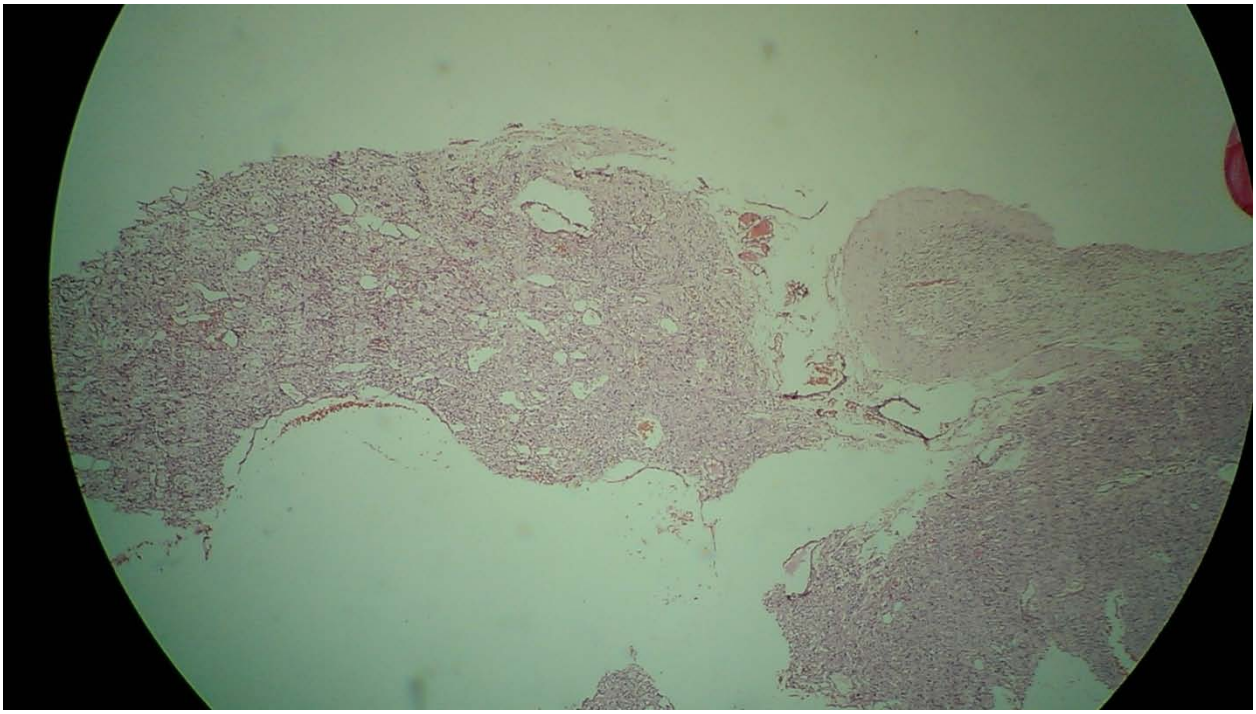


*Figure 2 :* HPE- Cerebellar Hemangioblastoma

Post operatively hemoglobin returned to 15gm%, patient was discharged with anti epileptics to review in 3 months.

## II. Discussion

Polycythemia refers to high hemoglobin greater than upper limit of normal, in adult females >16.5gm% or hematocrit >0.48, in adult males hemoglobin of 18.8gm% or hematocrit >0.52 due to increase number of red blood cells [true polycythemia] or reduction in blood volume [apparent, relative polycythemia].

Causes of true polycythemia are [1]primary polycythemia–a myeloproliferative disorder e.g. Polycythemia Rubra Vera; secondary true polycythemia due to increased secretion of erythropoietin secondary to tissue hypoxia as in high altitude, lung disease,

cyanotic heart disease, high affinity hemoglobin, or secondary polycythemia due to inappropriately increased erythropoietin secretion by renal diseases such as hydronephrosis, cysts, carcinoma, other tumors like bronchogenic carcinoma, uterine fibroids, hepatoma, pheochromocytoma,Cerebellar Hemangioblastoma.

Polycythemia Rubra Vera is a [2] clonal disorder involving a multipotent hematopoietic progenitor cell in which phenotypically normal red blood cells, granulocytes and platelets accumulate in the absence of a recognizable stimulus.

Since the patient had significant weight loss with true polycythemia in the second visit, not involving the other cells of granulocytes, and platelets, secondary polycythemia was considered; since ultra sonogram of the abdomen did not show any abnormality, search for tumors including Cerebellar Hemangioblastoma was undertaken.

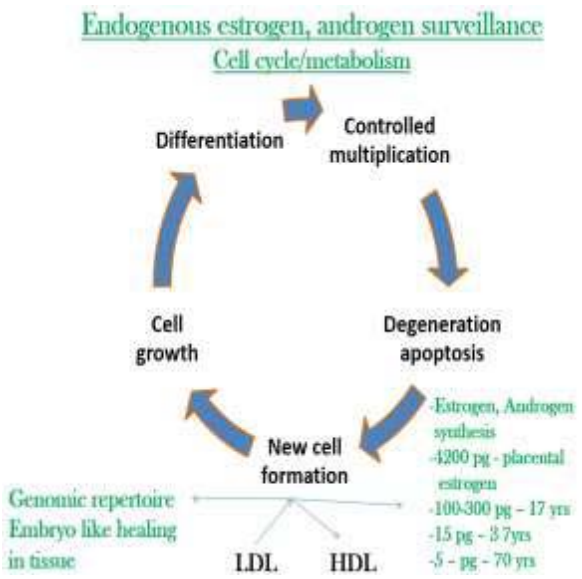
Hemangioblastoma are rare benign tumors that occupy central [3] nervous system and other regions like kidney, liver, pancreas; the incidence among posterior fossa tumors averages 11%; highest incidence is in 3rd to 6th decades of life; family history is significant with autosomal dominant inheritance, related to deletion of tumor suppressor gene, on the short arm of chromosome 3.

Hemangioblastoma are benign vascular tumors that arise from embryonic remnants of mesoderm origin, trapped in nervous system, during first trimester [4] of life Cerebellar Hemangioblastoma a rare cause of [5] erythrocytosis, consequent to ectopic production of erythropoietin by the tumor cells.

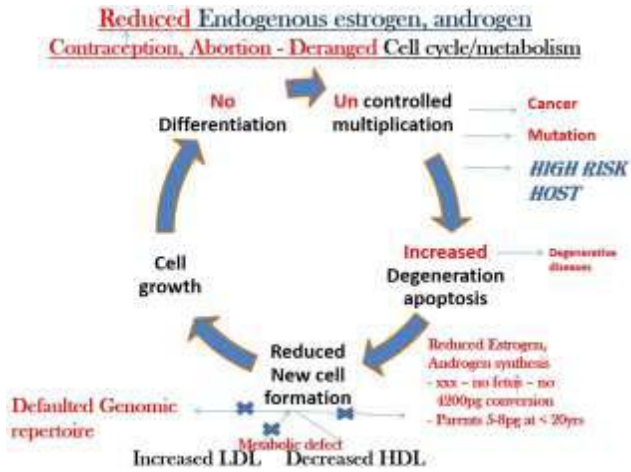
2 cases of Hemangioblastoma followed up long term showed malignant [6]behavior of transition to renal cell carcinoma from renal cysts

Hemangioblastoma is a tumor of the central nervous system, arising from vascular system, and can be present in retina [7 8 9 10] and spinal cord; can be associated with polycythemia, pancreatic cysts and Von Hippel Lindau syndrome.

As per the corresponding Authors earlier publications, couples observing contraception have 4-7 fold [11]increase in tumors, cancers, among 35- >50years of age, by fragmentation of the germ cells with associated decreased surveillance by estrogen, androgen [12]resulting in uncontrolled proliferation of any cell, preceded by no differentiation, resulting in neoplasms. If only the wife`s permanent sterilization can be reversed by tubal recanalization by awareness, financial aid, surgical expertise, the germ cells destruction will stop and the androgen, estrogen [13] will return to 79.9% of age normal, restoring genomic repertoire, cell cycle of differentiation, followed by controlled multiplication, so that recurrence and malignant transformation of the tumor will be curtailed.




Reference 12-Figure (a)




Reference 12-Figure (b)

## III. Conclusion

56 year old male presented with spurious or relative polycythemia suspect, later on progressed to true secondary polycythemia with significant weight loss, prompting a search for neoplasms; clinically he had subtle Cerebellar signs and imaging confirmed the diagnosis of Cerebellar Hemangioblastoma, with associated obstructive hydrocephalus; emergency ventriculo peritoneal shunt followed by excision of the tumor was performed and the histopathological evaluation confirmed WHO grade 1 Cerebellar Hemangioblastoma; hemoglobin normalized with excision of the tumor.

## IV. Acknowledgements

*The needful expert services of Neurosurgery, General pathology, Radiology colleagues is deeply acknowledged with gratitude.*

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