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2 **RESPONSE TO REVIEWER 1 COMMENTS**

3 **Q1 This manuscript reviews 2 cases of tuberous sclerosis presenting in children. Both children**
4 **had retinal hamartomas. There is a nice review of the types of retinal hamartomas. However, the**
5 **manuscript does not add to the current body of literature.**

6 **A1 This manuscript contains the review of 2 cases of TS. These rare cases have rarely been reported**
7 **from our subcontinent and it could distribute important knowledge to our colleagues of other departments**
8 **that meticulous eye examination can be very rewarding to establish the diagnosis whenever TS is**
9 **suspected.**

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11 **Q2 The introduction is too long for the purposes of the paper.**

12 **A2 The introduction has been shortened.**

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14 **Q3 The English and grammar are awkward and often incorrect.**

15 **A3 We belong to a poor but developing country where English is not our native language but we have**
16 **tried our best to correct the grammatical mistakes.**

17

18 **Q4 The referencing is also often poor.**

19 **A4 The referencing system has been improved.**

20

21 **Q5 There is no comment on whether this manuscript was reviewed by the IRB or whether the**
22 **families consented to be included in this publication. A face is shown and the eyes are not**
23 **blocked.**

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25 **A5 The consent from IRB and family was obtained and documented at the end of the manuscript..**
26 **The eyes are blocked and identity has not been disclosed.**

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31 **RESPONSE TO REVIEWER 2 COMMENTS**

32 **Q1 The clinical diagnosis of tuberous sclerosis was made before the ophthalmic examination.**

33 **A1** The diagnosis of tuberous sclerosis was only suspected in paediatric department and the
34 establishment of this clinical diagnosis was done after ocular evaluation.

35

36 **Q2 Presence of retinal astrocytoma is one of the major diagnostic criteria of tuberous sclerosis.**

37 **Considering there are no pathognomonic signs for the diagnosis. The authors seemed to be**
38 **considering only retinal astrocytoma as a diagnosis .**

39 **A2** Out of the 11 major and 9 minor diagnostic criterias of TS, eye findings contribute to 1 major (Multiple
40 retinal nodular hamartomas) and 1 minor criteria (Retinal achromic patch). And Definite diagnosis of TS is
41 done with Either two major features or one major feature plus two minor features and Probable diagnosis
42 of TS is done with One major plus one minor feature. Hence, we believe presence of retinal astrocytoma
43 can significantly aid in the diagnosis of TS in presence of other systemic features.

44 **Q3 How was the vision recorded in a mentally retarded child as 20/20.**

45 **A3** The vision in case 1 could be recorded with Snellen's chart as the child was a grade V student and
46 his IQ level was just subnormal i.e. gross mental retardation was not present

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48 **Q4 The differential diagnosis of the retinal lesions should be discussed .**

49 **A4** The differential diagnosis have been added in introduction and discussed in discussion part.

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51 **Q5 Was the astrocytoma calcified? Because diagnosing a non-calcified astrocytoma can be**
52 **challenging especially in a fairly pigmented individual**

53 **A5** The astrocytoma in case 1 was calcified and in case 2 was noncalcified but its diagnosis was not
54 challenging because the child is from South Asia and was heavily pigmented.

55

56 **Q6 Was fluorescein angiography done? Discuss the findings.**

57 **A6** Unfortunately, due to the subnormal mental status of these children, FFA was not attempted.

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59 **Q7 Was evaluation of lung done as it is of prognostic significance.**

60 **A7** Evaluation of all the systems including the lung was done in the paediatric department.

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66 **RESPONSE TO REVIEWER 3 COMMENTS**

67 **Q1 The manuscript should have been revised by a native speaker; actually, it contains an**
68 **excessive number of errors.**

69 **A1 Report of Tuberous Sclerosis from developing countries like ours have rarely been done. Hence we**
70 **found it very worthful to report. It has been revised by a native speaker and the correction of the errors**
71 **has be done/**

72 **Q2 The introduction is too long and the discussion too short, and both contain some**
73 **inappropriate sentences**

74 **A2 The introduction is shorten, discussion is lengthen and inappropriate sentences removed.**

75 **Q3 Abstract: conclusion: it seems more appropriate to affirm that retinal phakomatosis aids in**
76 **confirming TSC instead of “ruling out”.**

77 **A3 Correction done**

78 **Q4 Introduction/Discussion: line 38: the so called SEGAs (subependymal giant astrocytomas)**
79 **have to be large at least 10 mm... the author probably means subependymal periventricular**
80 **nodules.Lines 59-62: this sentence is inappropriate, it doesn't add anything useful.Line 66-68: it**
81 **seems more appropriate to move it to the discussion.**

82 **A4 Corrections done.**

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91 **RESPONSE TO REVIEWER 4 COMMENTS**

92 **Q1 Line 31 `Worthful' - to be replaced by `essential**

93 **A1 Replaced**

94 **Q2 Line 37 `hamartias' – to be deleted.**

95 **A2 Deleted**

96 **Q3 Line 59-62 `vitreous----- haemorrhage.' – sentence may be deleted.**

97 **A3 Deleted**

98 **Q4 Line 79- supporting investigation reports must be stated.**

99 **A4 Added**

100 **Q5 Line 82 `speaking' – to be replaced by speech.**

101 **A5 Replaced**

102 **Q6 Line 106 `in he family' – to be replaced by `in the family'**

103 **A6 Replaced**

104 **Q7 Line 135 `under clonazepam' – to be replaced by under treatment of clonazepam.**

105 **A7 Replaced**

106 **Q8 Line 153 `ia reported – to be replaced by have been.**

107 **A8 Replaced**

108 **Q9 Line 161 `child of seizure with skin lesion' – to be replaced by `child with seizure and skin**

109 **lesions**

110 **A9 Replaced**

111 **Q10 The case 1 had sebaceous adenoma and the case 2 had angiofibromatosis ,**

112 **hisopathological findingd/histopathological pictures of both should be provided.**

113 **A10 Unfortunately,the histopathological findings of the skin lesions were not available to us from the**

114 **dermatology department**

115 **Q11 Identity of child in photograph should not be revealed. Black tape may be put on eyes**

116 **A11 Identity of child in photograph is not revealed by putting a black tape on eyes.**

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119 **RESPONSE TO REVIEWER 5 COMMENTS**

120 **Compulsory REVISION comments**

121 **Q1The introduction is too long . For example lines 40-46 and 54-56 can be extremely**
122 **shortened and even omitted. Other irrelevant data concerning this specific article should**
123 **also be omitted .**

124 **A1.Introduction has been shortened by omitting the irrelevant data.**

125 **Q2 Lines 86 and 121. Negative for what? TS?**

126 **A2 Lines 86 and 121 meant no similar history was present in the family and these lines have now**
127 **been removed.**

128 **Q3 Giving the IOP, axial length and retinscopy findings adds no relevant information to**
129 **this paper.**

130 **A3 IOP, axial length and retinscopy findings were mentioned to support the fact that the eyes were**
131 **otherwise healthy.**

132 **Q4 What about US? OCT? If done should be added.**

133 **A4 The US was done and findings has been documented, unfortunately,OCT was not done.**

134 **Q5 The MRI's are very small and one cannot exactly identify the lesions mentioned in the**
135 **legend, Arrows should be added.**

136 **A5 The MRI has been enlarged and arrows has been added to show MRI lesions.**

137

138 **Q6 This is a study describes 2 children with TS. The figures are very illustrative but I**
139 **don't recognize any new important information this paper adds . Retinal astrocytomas are**
140 **a known ocular finding in patients with TS.**

141 **A6 This manuscript contains the review of 2 cases of TS.These rare cases have rarely been**
142 **reported from our subcontinent and it could distribute important knowledge to our colleagues of other**
143 **departments that meticulous eye examination can be very rewarding to establish the diagnosis whenever**

144 TS is suspected because retinal astrocytoma is 1 one of the 11 major and 9 minor diagnostic criterias of
145 TS.

146 **Minor REVISION comments**

147 **Q7 The figures are very nice. I think figures 1+5 should be united and also 2+3+6**

148 **A7 The figures 1,2,3,4 are of case 1 and 5,6 belong to case 2,hence they are shown separately.**

149 **Q8 OPD (line 76) the abb. Should be explained.**

150 **A8 The word OPD means outpatient department and has been removed from the text.**

151 **Optional/General comments**

152 **Q9 This paper suffers from many serious language issues (too many to be mentioned) and**
153 **should be edited**

154 **A9 Edition done.**

155 **Q10 Figure 2 contain a full face photograph of a child. The eyes should be hidden since the**
156 **main finding is on the skin.**

157 **A10 The identity of the child has been hidden by putting a black tape over the eyes.**

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159 **RESPONSE TO REVIEWER 6 COMMENTS**

160 **Q It is a rare entity described, documentation is good enough. English language is sufficient. I**
161 **would accept it without revision.**

162 **A Thank you.**

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